



Fig. 2.
Placental capillaries. (a) Part of the film on capillary action in the placenta, before adding histamine. (b) After adding histamine the capillaries are dilated.

1954), which fill the blood-vessels of the villi more or less intermittently, so that the volume varies from one moment to the next. This affects the width of the narrow crevices around the villi. Thus maternal blood in these crevices, which form part of the intervillous space, circulates by the action of foetal pre-capillaries in the placenta (Fig. 1). The arteries in the basal layer of the placenta dilate before entering the intervillous space, and pressure in them is low; the veins and arteries open close

together in the basal layers of the placenta, so that arterial blood could pass easily to the veins. But, by suction and circulation, blood is transported to the centre of the placental bed. There are fewer villi at the edges of the placenta and here the venous return to the large uterine veins is not impeded. The action of the pre-capillaries in the placental villi is automatic and can be observed after the expulsion of the placenta. We have made a film to illustrate this phenomenon.

TABLE I—PERINATAL MORTALITY IN CASES OF TOXAEMIA TREATED BY INDUCTION OF LABOUR

Number of toxæmic patients	294
Number on whom oestriol determinations (2) were performed	104
	<i>Perinatal Deaths</i>
All toxæmic cases	27 (9%)
Induction of labour after oestriol assays	14 (14%)
Malformation	2
Hydrops	1
Toxaemia only	11 (10%)
Under 34 weeks maturity	5
Hospital admission too late	1
Induction too late	7 (7%)
Total	27

TABLE II—FOETAL DEATHS IN CASES OF TOXAEMIA OF PREGNANCY IN WHICH SERIAL OESTRIOL ASSAYS WERE CARRIED OUT

Case No.	Parity	Cause of Foetal Death	Gestation (weeks)	Oestriol Assay (γ)
1	Ip. (0)*	Pre-eclampsia, induction too late	36	< 10,000
2	IIp. (3)	Hypertensive disease, death <i>in utero</i> , induction too late	35	< 10,000
3	IIIp. (1)	Hypertensive disease, death <i>in utero</i> , induction too late	36	< 10,000
4	IIp. (0)	Hypertensive disease, macerated foetus, malformation	36	< 10,000
5	Vp. (0)	Vaginal bleeding, dead foetus	26	< 1,000
6	Ip. (0)	Pre-eclampsia, death <i>in utero</i> , induction too late ..	36	< 10,000
7	IIIp. (0)	Hypertensive disease, death <i>in utero</i> , induction too late	36	< 10,000
8	VIIIp. (2)	Hypertensive disease, Rh. hydrops	40	< 10,000
9	VIIp. (0)	Hypertensive disease, death <i>in utero</i>	28	< 10,000
10	VIp. (0)	Hypertensive disease, death <i>in utero</i> , too premature ..	34	< 10,000
11	IVp. (0)	Pre-eclampsia, death <i>post partum</i> , severe malformation	40	\pm 18,000
12	Ip. (0)	Pre-eclampsia, death <i>in utero</i> , induction too late ..	40	> 12,600
13	Vp. (0)	Pre-eclampsia, death <i>in utero</i> , admission too late ..	36	< 10,000
14	IIIp. (0)	Pre-eclampsia, death <i>in utero</i> , induction too late ..	37	< 12,500

* Numerals in parentheses are abortions

TABLE III—DIABETICS. OESTRIOL VALUES IN RELATION TO PLACENTAL ABNORMALITIES

Case	Control of Diabetes	Weight of Child ÷ Weight of Placenta	Oestriol		Number of Abnormal Features in Microscopical Examination of Placenta (Total 7 Features)	Week of Gestation
			Main Excretion	Last Value before Delivery (Y) (weeks)		
A	Good	$\frac{3970}{550} = 7.2$	Increased	33,100 40	6/7	40
B	Irregular	$\frac{3190}{375} = 8.5$	Value only after death of child increased before death of child		4/7	†
C	Irregular	$\frac{4300}{398} = 10.8$		32,200 36	5/7	†
D	Rather irregular	$\frac{4050}{600} = 6.6$	Increased	33,100 39	2/7	39
E	Good	$\frac{3450}{450} = 7.6$	Increased	46,000 39	1/7	40
F	Good	$\frac{3360}{?} = ?$	Decreased	26,300 39	—	39
G	Good	$\frac{3220}{700} = 4.6$	Decreased	18,100 36	3/7	36
H	Good	$\frac{3480}{650} = 5.3$	Decreased	20,000 40	5/7	40
I	Good	$\frac{3460}{750} = 4.6$	Decreased	8,000 35	—	36
J	Good	$\frac{4900}{630} = 7.7$	Decreased	10,000 36	—	± 36
K	Good	$\frac{3640}{640} = 5.7$	Decreased	17,600 36	7/7	36
L	Good	$\frac{2840}{420} = 6.7$	Decreased	11,500 36	—	36
M	Irregular	$\frac{?}{750} = ?$	Decreased	5,100 38	6/7	38

† = Post-natal

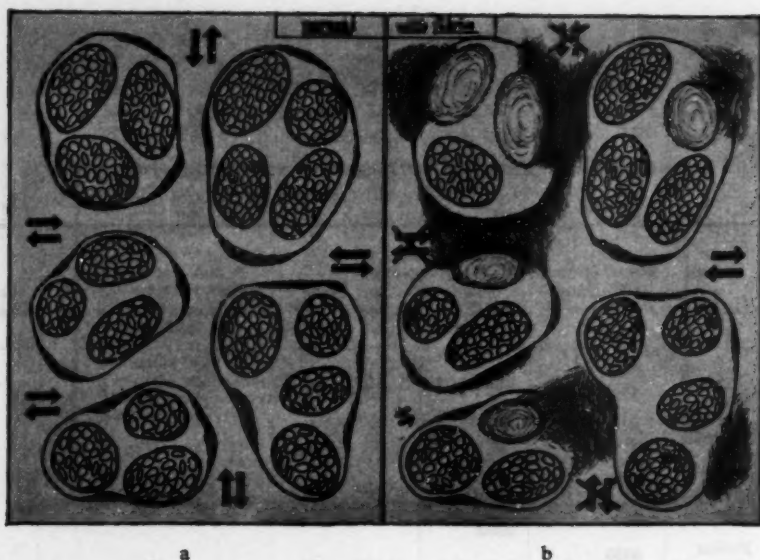


Fig. 3. Effect of fibrin precipitation in intervillous space from thrombosis of foetal vessels. Reduced capillary action and degeneration of epithelium. (a) Normal. (b) Precipitation of fibrin.

Capillary action is affected by substances such as histamine (Fig. 2a and b), acetylcholine and progesterone. The pre-capillaries in the placental bed affect the oxygenation of the foetus. This influence is less in toxæmic placentas, in which more capillaries are closed by thrombi, with precipitation of fibrin around the villi causing them to adhere to one another (Fig. 3a and b). These changes are also found in placentas at term and later, and are an indication of postmaturity.

The foetal vessels in the villi of diabetic and syphilitic placentas and in cases of rhesus incompatibility are small and contracted and situated in the centre of the villi, and the epithelium of the villi is thickened. It is known that foetal oxygenation is disturbed in these conditions and the children have low oxygen reserves. Death *in utero* is common in toxæmia, diabetes, syphilis and rhesus incompatibility because of placental insufficiency.

By serial urinary oestriol determinations we have been able to measure placental insufficiency in hypertensive toxæmia and prevent intra-uterine death by induction of labour at the optimum time (ten Berge 1957). From 1956 to 1958, 104 patients with severe toxæmia were investigated for diminished oestriol excretion, indicating placental degeneration in the last 8 weeks. Figures below 10,000 gamma oestriol per 24 hours indicate danger to foetal life and the advisability of Caesarian section. Results are shown in Tables I and II.

We found a sudden fall in urinary oestriol within 24 hours in cases of sudden foetal death (Fig. 4). We attributed this to cessation of capillary pulsation with an abrupt decrease in blood-flow through the intervillous spaces causing diminished transport of oestriol (ten Berge 1960a).

Hormonal production continues for some time after foetal death, as may be observed from the level of urinary

oestriol, which although low continues until degeneration of chorionic epithelium progresses and oestriol production ceases entirely. The same phenomenon was observed in cases of placenta praevia, where localisation of the placenta with 5 microcuries of intravenous radioactive iodinated human serum albumen was carried out (ten Berge 1960b). A dead foetus could not be localised on account of the low blood-circulation in the intervillous spaces.

Determination of oestriol values is one example of improved antenatal care, but is only of use in hypertensive toxæmia of pregnancy. The method was found unsuitable in diabetes. Here microscopical examination of placentas showed:

1. Large villi.
2. Oedema.
3. Hyperplasia of stroma.
4. Presence of Hofbauer cells.
5. Small blood-vessels in the central stroma of the villi.
6. Epithelial proliferation.
7. Presence of Langhans cells.

We also studied the weight of the placentas in relation to the weight of the child. We compared the index:

Weight of child/Weight of placenta

with the oestriol values. The figures are shown in Table III. It may be concluded that in diabetes there is no connection between urinary oestriol and the condition of either the placenta or the foetus. Placental changes in diabetes differ from those which occur in toxæmia and vary from case to case. The indications for induction of labour in diabetes are the onset of hydramnios, increasing weight of the baby, and the progress of the disease. Postmaturity may be a further indication, as judged by examination of vaginal smears.

Our methods of determining placental function and of assessing the condition of the baby are still incomplete. Our task is to improve these methods and find a safe method for the induction of labour. We recommend:

1. That oxygen should be given in labour, particularly if this is prolonged.
2. That we should try to estimate placental function.
3. That expulsion should be hastened by relaxation of the levator muscles of the pelvic floor.
4. That use of the vacuum extractor or forceps should be avoided where possible.
5. In breech presentations Bracht's method (spontaneous expulsion with applied expression) should be employed.
6. The avoidance of anaesthesia. If the mother is psychically prepared she will be in a state of tranquillity and will not require anaesthesia. Attention should be paid to her respiration and the oxygenation of the child.
7. Recording of the foetal heart in preference to auscultation, so that early disturbances may be recorded and action taken to prevent foetal distress.
8. Regulation of labour pains in conjunction with foetal heart action. Good drugs for this purpose are lacking.

The application of more efficient measures will be amply rewarded by the reduction of human suffering and the preservation of the children's intelligence. The obstetrician who makes this his task should restrict his work entirely to obstetrics. The problems of pregnancy and obstetrics demand the setting up of special obstetric biochemical and physiological laboratories.

SUMMARY

Former studies of cerebral anomalies have mostly been concentrated on the effects of parturition, using retrospective investigations. Since 1954 prospective investigations have been carried out in Groningen, using the methods devised by Dr. Heinz Prechtel, with follow-up studies by Dr. J. Dijkstra.

Latent oxygen deficiency in the last weeks of pregnancy is as important in the causation of cerebral defects as damage incurred during labour. Special attention has been paid to the anatomy and functions of the placenta and the foetal circulation of the villi. The circulation of the mother's blood in the intervillous space depends on the action of the pre-capillaries and capillaries in the villi. This action is independent and continues after expulsion of the placenta. Circulatory disturbances are described in cases of vascular toxæmia, diabetes, rhesus incompatibility, etc.

In future, obstetricians should pay more attention to the degree of placental damage and the compensatory mechanisms. Here, serial oestrogen estimations in 24-hour urine have proved of great value in vascular toxæmia. In these cases they enable the induction of labour to be timed more accurately. In diabetes the tests have no particular value and clinical signs are more important. Localisation of the placenta by isotope methods is of value in cases of bleeding in the last three months of pregnancy.

Another task for future inquiries is to find a safe and efficient method for the induction of labour which will take account of the child's condition in utero and regulate uterine contractions during labour accordingly. Recommendations are made for dealing with conditions where the foetal oxygen reserves are low.

RÉSUMÉ

Les études antérieures sur les anomalies du cerveau ont porté, dans la plupart des cas, sur les effets de la parturition au moyen d'investigations retrospectives. Depuis 1954, des recherches projectives ont été effectuées à Groningen à l'aide des méthodes mises au point par le Dr. Heinz Prechtel et des recherches complémentaires du Dr. J. Dijkstra.

Il a été établi que la déficience latente en oxygène au cours des dernières semaines de la grossesse était un facteur causal de malformations cérébrales aussi important que les lésions subies pendant le travail.

L'anatomie et les fonctions du placenta et de la circulation foetale dans les villosités ont particulièrement retenu l'attention. La circulation du sang de la mère dans l'espace villositaire dépend de l'action des précapillaires et des capillaires dans les villosités. Cette action est indépendante et se prolonge après l'expulsion du placenta. Les troubles circulatoires sont décrits dans la toxémie vasculaire, le diabète, l'antagonisme rhesus, etc.

Désormais les accoucheurs devront prêter plus d'attention au degré de dégradation du placenta et des mécanismes de compensation. A cet effet, des évaluations en série d'oestriol sur des urines de 24h. ont avéré leur grande efficacité dans la toxémie vasculaire. Dans ces cas, ils permettent d'induire le moment du travail avec plus de précision. Dans le diabète, les évaluations n'ont pas beaucoup d'intérêt et les signes cliniques sont plus significatifs. La localisation du placenta par les isotopes radioactifs est d'un grand intérêt dans les cas d'hémorragies au cours des trois dernières semaines de la grossesse.

Il reste encore à trouver une méthode sûre et efficace pour l'induction du travail qui tienne compte de l'état de l'enfant in utero, tout en régularisant les contractions utérines pendant le travail.

Il est fait certaines recommandations dans le cas où les réserves en oxygène foetal sont faibles.

ZUSAMMENFASSUNG

Die früheren Studien über Gehirnanomalieen bezogen sich meistens auf die Folgen der Geburt, mittels retrospektiver Forschungen. Seit 1954 sind prospektive Forschungen in Gröningen durchgeführt worden, mittels der von Dr. Heinz Prechtel erfundenen Methoden und der Vervollständigungen Dr. J. Dijkstra's.

Es wurde festgestellt, dass latenter Sauerstoffmangel in den letzten Wochen der Schwangerschaft eine ebenso wichtige Ursache der zerebralen Defekte ist, wie während der Geburt erlittener Schaden. Besondere Aufmerksamkeit ist der Anatomie und den Funktionen des Plazentas und des foetalen Kreislaufs in den Zotten gewidmet worden. Der Kreislauf des mütterlichen Blutes im intervillösen Raum hängt von der Tätigkeit der Präkapillaren und Kapillaren in den Zotten ab. Diese Tätigkeit ist selbständig und setzt nach Ausstossung des Plazentas fort. Kreislaufstörungen werden bei Gefäßtoxämie, Diabetes, Rhesusantagonismus u.s.w. beschrieben.

In der Zukunft müssen die Geburtshelfer mehr auf den Grad der Schädigung des Plazentas und der Prophylaxis achten. In dieser Absicht haben sich Reihenabschätzungen des Oestriols in 24 stündigem Harn sehr wertvoll bei vaskulärer Toxämie erwiesen. In diesen Fällen geben sie die Möglichkeit, die Induktion der Geburtswehen genauer zu regeln. Bei Diabetes besitzen die Prüfungen keinen besonderen Wert und klinische Symptome sind wichtiger. Lokalisierung des Plazentas durch Isotopmethoden hat grossen Wert bei den Fällen von Blutungen in den letzten drei Monaten der Schwangerschaft.

Eine andere Aufgabe für zukünftige Forschungen besteht darin, eine sichere und wirksame Methode für die Induktion der Entbindung zu finden, die den Zustand des Kindes in utero in Rechnung zieht und die Zusammenziehungen der Gebärmutter während der Entbindung demgemäss reguliert.

Es werden Empfehlungen gemacht für die Fälle, wo die foetalen Sauerstoffreserven gering sind.

REFERENCES

- ten Berge, B. S. (1955a) 'De bloedstroom in de placentavlokken.' *Ned. T. Geneesk.*, **99**, 2371-2377.
- (1955b) 'The bloodstream through the placental villi.' *Int. J. Fertility*, **1**, 31.
- (1955c) *Tendances actuelles en Gynécologie et Obstétrique*. Geneva: Georg & Cie. (Film).
- (1956) 'L'activité capillaire dans les villosités placentaires.' *Bull. Soc. roy. belge Gynéc. Obstét.*
- (1957) 'Oestriol assays during pregnancy.' *Acta endocr. (Kbh.)*, suppl. 31, 31.
- (1959) 'Placenta and oestriol excretion in diabetes during pregnancy.' In Symposium on Prenatal Care. pp. 160-167. Groningen: Noordhoff.
- (1960a) 'Oestriol excretion in intra-uterine foetal death: the significance of a sudden decrease and its relation to capillary pressure in the villi.' *Gynaecologia*, **149**, 40-55.
- (1960b) 'Placentallocalization mittels Röntgen und Isotopen.' *Berl. Med.* 491.
- Bøe, F. (1954) 'Vascular morphology of the human placenta.' In *Cold Spr. Harb. Symp. quant. Biol.* **19**, pp. 29-35.
- Dijkstra, J. (1960) 'De prognostische betekenis van neurologische afwijkingen bij pasgeboren kinderen.' Thesis. Groningen.
- Eastman, N. J. (1954) 'Mount Everest in utero.' *Amer. J. Obstet. Gynec.*, **67**, 701.
- Little, W. J. (1862) 'On the influence of abnormal parturition, difficult labour, premature birth and asphyxia on the mental and physical condition of the child, especially in relation to deformities.' *Trans. obstet. Soc. Lond.*, **3**, 293. Reprinted in *Cer. Palsy Bull.* Vol. 1, no. 1, 1958, pp. 5-36.
- Prechtel, H. F. R. (1958) 'The directed head turning response and allied movements of the human baby.' *Behavior*, **13**, 212.
- Walker, J. (1954) 'Foetal anoxia. A clinical and laboratory study.' *J. Obstet. Gynaec. Brit. Emp.*, **61**, 162.

NOTICE**Mental Health Book Review Index**

THE *Mental Health Book Review Index*^{1,2}—lists references to signed book reviews appearing since January 1955 in 132 journals in the English language. No book is included unless it has been reviewed in at least three journals, but according to the compilers neither inclusion nor omission of a book implies a definitive selection. The *Index* is sponsored by the World Federation for Mental Health, the American Foundation for Mental Hygiene, and other bodies. Its compilers, Ilse Bry and Margaret Kinney, are well known for their work on the indexing of the literature of psychiatry and psychology.

A paper³ written by Miss Kinney, Miss Franck and Dr. Bry in 1955 gives some interesting observations on book reviews in general. Previous writings on the subject are dismissed rather lightly and it is suggested that little systematic research has been done in this field. One doubts whether there is in fact such a paucity of literature. To take one example only, the controversial question whether reviews should or should not be signed has received considerable attention in British medical journals.

Dr. Bry and Miss Kinney say that the *Index* has demonstrated that the number of reviews is no measure of the value of a book—'furthermore we now know that scholarly exchanges of reactions to a book can last as long as five years.' One might have thought that these conclusions could have been reached by a less laborious method than the listing of 8,500 citations of reviews. What the editors are apparently seeking is 'an objective method of selecting books in the behavioral sciences and mental health, and a new way of organising this literature.' It is, however, admitted that the books listed in the first ten issues of the *Index* 'are neither a selection of outstanding contributions nor a representative sample of the recent behavioral-science literature.' The *Index* represents an enormous amount of work and it should be of value to those concerned with the widely dispersed literature in this field. At the same time one cannot help wondering whether a listing of the same books under subject headings, with very brief evaluative notes, would not prove even more useful. Such a guide could almost certainly be produced with much less labour and expense. The compilers express the hope that they will eventually 'be able to show not only that the mills of the scientific community grind slowly, but also that they grind exceeding small.' Is it possible that they sometimes grind too small?

W. J. Bishop

¹ *Mental Health Book Review Index*. Vol. 5, No. 1, January 1960; Vol. 5, No. 2, July 1960. Two issues per year—\$3.00. Single issues—\$2.00 each, obtainable from Miss Lois Afflerbach, Paul Klapper Library, Queens College, Flushing 67, N.Y.

² Editorial. '*Mental Health Book Review Index*'. *A.M.A. Archives of General Psychiatry*, 1960, 2, pp. 701-706.

³ Kinney, M. M., Franck, M. and Bry, I. The Book Review—A Hybrid in Literature and a Stepchild in Documentation. Reprint from International Congress of Libraries and Documentation Centers, Brussels, 1955, Vol. II A, Communications.

Palm-prints and their Uses in Medical Biology

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RIDGED skin occurs on definite regions of the human body; on the palmar surface of hands and fingers and on the plantar surface of feet and toes. The epidermal ridges and their arrangements are studied by taking prints of the areas concerned. Prints of palms and finger tips can be taken with little difficulty and supply permanent records of the ridge configurations in these areas. Sole-prints, particularly of adults, cannot always be obtained so readily, while the printing of toes presents certain technical difficulties. Hence, our knowledge of the ridged skin on palms and fingers is much greater than that on soles and toes.

The use of finger-prints and, to a lesser extent, palm-prints and sole-prints, for personal identification is well known and has attracted much interest. It is, however, often supposed, mistakenly, that the study of epidermal ridges (dermatoglyphics*) is unimportant, save for this purpose. Applications in human biology are of a more fundamental nature. Palm-prints and finger-prints have been widely used in twin studies, chiefly in the diagnosis of zygotic type. For some time it has been realised that growth disturbances in foetal life cause modifications of the ridge arrangements. Palmar patterns are sensitive indicators of developmental anomalies.

* Until 1926 there was no term to embrace the study of ridged skin on fingers, toes, palms and soles. Cummins and Midlo (1926) proposed the word dermatoglyphics (*derma*, skin and *glyphein*, to carve) as a collective name for all the features of ridged skin and also for the study of dermal ridges and their arrangements. The term is now in general use among biological workers in this field.

Moreover, it is now known that ridge arrangements show significant distortion in cases where chromosome abnormalities are present.

Characteristics of Dermal Ridges

To understand the value of dermal ridge impressions it is necessary to know the principal characteristics of ridged skin. Unlike most human traits, dermal ridges and the configurations formed by them are age-stable. Ridge differentiation takes place in the foetus during the third and fourth pre-natal months. *By the end of the fourth foetal month, the ridges and patterns (designs composed of markedly curved ridges) are complete and in their permanent form.* After this they do not change morphologically throughout life and indeed they persist for some time after death. Growth in size keeps pace with the growth of hands and feet, but the detailed structure of the ridges and their arrangement remain the same. Environmental factors can only affect ridge-formation in the uterus and from birth dermatoglyphic features are environment-stable.

The detailed structure of individual dermal ridges varies greatly. The details of a small area of ridged skin are not repeated either in the same or a different individual. Even the dermatoglyphic features on the hands of monozygotic twins are not identical, although the likenesses are remarkable. Neither are the two palms of one person ever exactly alike. The pattern-



Plate 1A. Photograph of a plaster cast of the left palm of an adult male showing the arrangement of dermal ridges.



1B. Palm-print of the same hand as in A. Note that it is a mirror-image of the cast. The impressions of the finger patterns are incomplete and are called 'plain' impressions. Unless the finger patterns are small, 'rolled' prints of each finger are necessary to obtain complete impressions of the patterns.

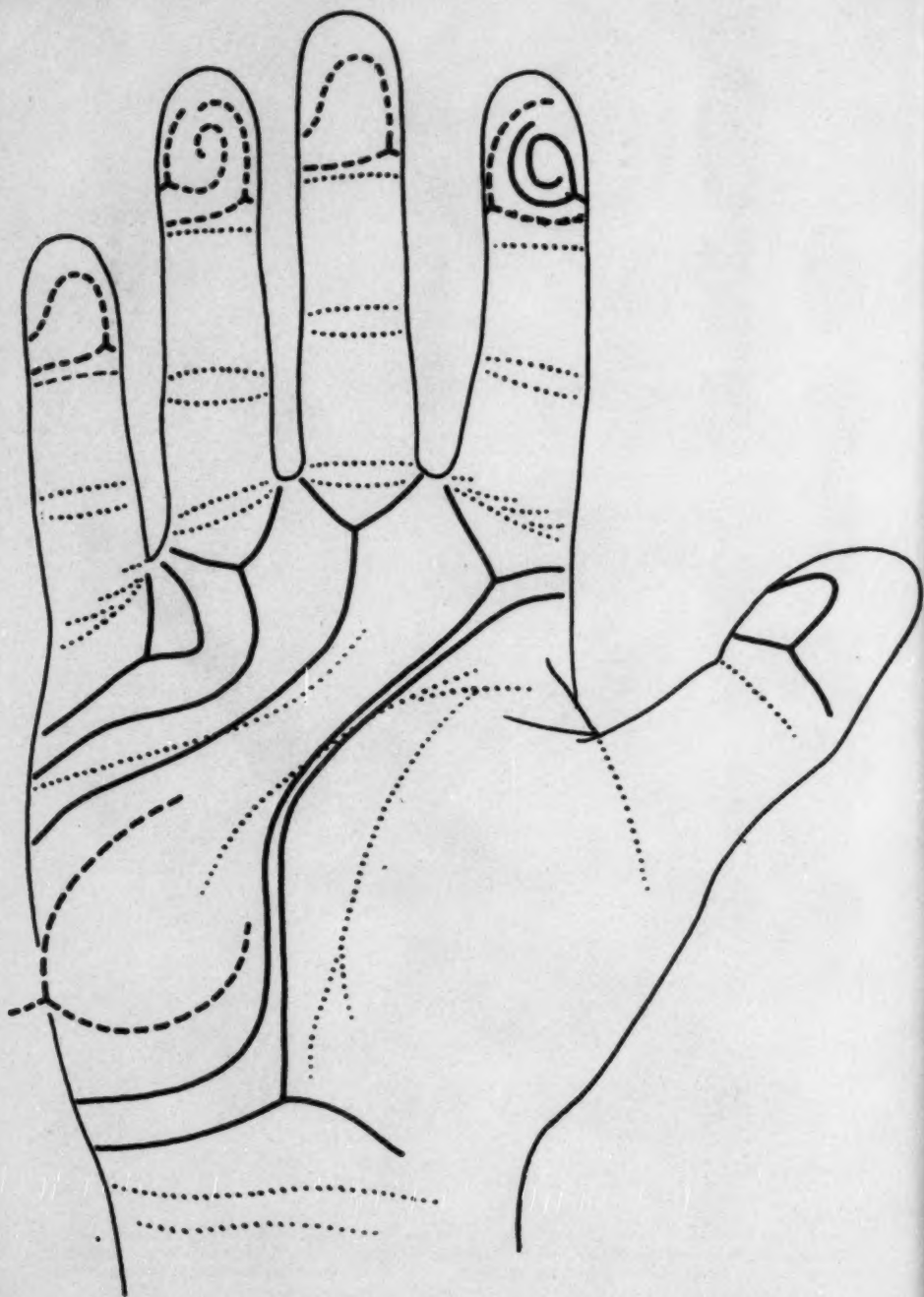


Fig. 2. Tracing of palm-print in plate 1B, showing the main lines and chief features of the ridge arrangement. The type-lines of the thumb have been traced from a rolled finger-print.

types, however, though very variable, vary within limits, so that classification is possible.

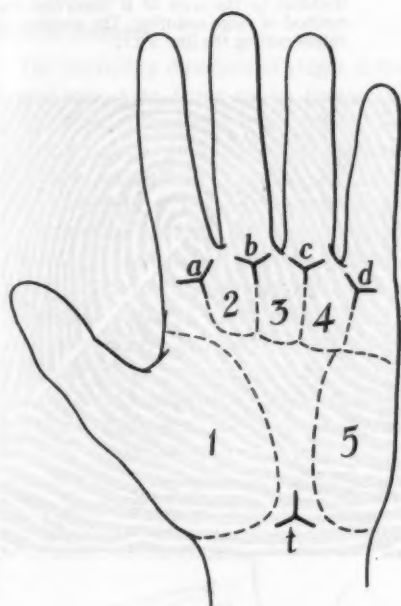


Fig. 1. Diagram of a typical right hand showing the position of the principal palmar triradii and the five areas of the palm where patterns may occur. *a*, *b*, *c* and *d* are the digital triradii and *t* the axial triradius. 1: Thenar/first interdigital area. 2: Second interdigital area. 3: Third interdigital area. 4: Fourth interdigital area. 5: Hypothenar area.

General Features of Ridge Arrangement on Palms

On examining a palm-print (see Plate 1B) it can be seen that the skin ridges course in different directions in different areas of the palm. Where three ridge systems meet the ridges form a triradiate structure, the *triradius*. Typically a triradius occurs at the base of each digit except the thumb. These are called the *digital triradii* and designated *a*, *b*, *c* and *d*. Triradius *a* is situated in the distal palm under the index finger, *b*, *c* and

d, in that order, under the middle finger, ring finger and little finger respectively. Another triradius is usually found at the base of the palm and is called the *axial triradius*, *t*. The positions of the five principal triradii of the palm are shown in Fig. 1.

Other triradii are associated with patterns. By marking on a palm-print, with a fine-pointed HB pencil, the ridges (or radiants) which issue from each triradius and tracing the course of these ridges across the palm, the chief features of the ridge arrangement are shown up. In order that tracings should be accurate various rules have been laid down and these are given by Cummins and Midlo (1943).

The tracings from the radiants of *a*, *b*, *c*, *d* and *t* that are directed to the centre of the palm are called the *main-lines* of the hand. A tracing of the main-lines of the palm of an adult male is shown in Fig. 2 and another in Fig. 5A. The terminations of the main-lines vary to some extent from palm to palm, though within certain limits.

In definite areas the ridges may be arranged to form patterns. There are five of these areas on the palm: the *thenar/first interdigital*, the *second*, *third* and *fourth interdigital* and the *hypothenar* (see Fig. 1). In addition, patterns occur on each of the ten finger-tips. The outlines or *type-lines* of finger patterns can be traced from the triradii in a similar manner to that used for palmar main-lines and patterns. The ridge configurations are very variable and are determined partly by heredity and partly by such factors as stress and tension in the growth of the part during foetal life.

Finger Patterns

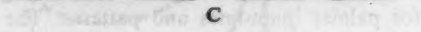
Finger-print patterns were classified by Galton (1892) into three main types—*arches*, *loops* and *whorls*—according to the number of triradii present. A simple arch has no triradius and is not a pattern at all



A



B



C

Fig. 3. Finger-prints showing the three main pattern-types. A: Arch (no triradius). B: Loop (1 triradius). C: Whorl (2 triradii). Note the increasing complexity of the patterns. The white line drawn from the triradius to the core of B illustrates the method of ridge-counting. The number of ridges cutting the line is 15.

in the true sense, being composed of a succession of gently curving ridges. A loop has one triradius and in a whorl there are typically two triradii (see Fig. 3). Loops are of two sorts, according to the direction they face. An *ulnar* loop opens towards the ulnar margin of the hand and has the triradius on the radial side, while a *radial* loop opens towards the radial margin and has the triradius on the ulnar side.

Patterns vary from digit to digit of an

individual. One person may have the same type of pattern on all ten fingers or various patterns may occur on different digits. Patterns vary in size as well as in shape. Loops are the commonest type and represent 70 per cent of all finger-patterns in Britain. The following are the frequencies of pattern types on fingers found in the British population:

Whorls	25%
Loops	{ Ulnar 64% Radial 6% }	.. 70%
Arches	5%

Certain patterns tend to occur more frequently on some fingers than on others. Thus whorls have their maximum frequency on digits I (thumb) and IV (ring finger), while radial loops and arches are most commonly found on digit II (index finger).

Digit V (little finger) has the highest frequency of ulnar loops and the lowest frequency of other patterns.

Palmar Patterns

The prevailing direction of ridges in the hypothenar area tends to be transverse, while in the thenar area it is vertical. The principle types of pattern in the hypothenar area are loops and whorls, the latter often having three triradii. Double-looped arrangements forming S-shaped patterns also occur. Interdigital patterns generally consist of loops opening into the nearest interdigital interval, but small whorls occur very rarely in these areas. Thenar/first interdigital patterns usually have a characteristic appearance with ridges running at right angles to the general direction (see Fig. 4). Whorls are, however, sometimes found in this part of the palm, but are much rarer.

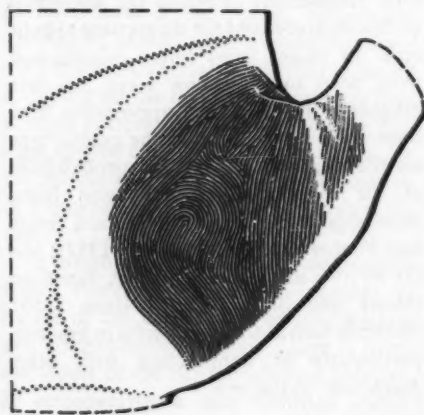


Fig. 4. A common type of thenar/first interdigital pattern.

Bimanual Differences

It has already been stated that the two palms of one individual are never exactly alike. Dermatoglyphic studies show that

dermal ridges present bilateral asymmetry in ridge breadth, pattern-size and pattern intensity. There is a tendency for patterns to be larger on right hands. On palms hypothenar, second and third interdigital patterns have higher frequencies in right hands, while thenar and fourth interdigital patterns are more common in left hands. On fingers, ridges are finer in left hands. As on palms, differential trends of pattern type are noticeable; symmetrical whorls and radial loops are more frequent in right hands and ulnar loops and arches in left hands.

Sex and Racial Differences

In a similar way, the sexes show certain dermatoglyphic differences. Ridges are on the whole wider in males than in females. Females have more arches and usually a lower frequency of whorls on their fingers, while on palms the frequencies of fourth interdigital and hypothenar patterns are higher. Males show more thenar/first interdigital and second and third interdigital patterns. Bimanual differences tend to be rather less marked on both fingers and palms in females than in males.

Racial differences are also evident. These show chiefly in differential frequencies of various pattern-types, both digital and palmar. For instance, many Asiatic populations have a much higher percentage of finger whorls than is found in the British population. It is not possible to tell from dermal prints to which race any particular individual belongs, for no pattern is characteristic of any race. In Europe pattern intensity tends to increase from north to south, while in Asia, Africa and in the indigenous populations of the Americas the reverse is roughly true.

It will be seen from these considerations that for dermatoglyphic studies samples must be carefully chosen and should be as homogeneous as possible racially, while the sexes need to be treated separately.

Inheritance

There is no doubt that many dermatoglyphic traits have a hereditary basis. This is shown by the marked similarities found in the palm- and finger-prints of monozygotic twins, while those of dizygotic twins are no more alike than the prints of ordinary sibs. The genetic processes involved are complex and, though much work has been done on the subject, we do not yet know how the pattern-types are inherited. In recent years, however, quantitative methods have been applied. By measurement of the angle maximal *and* on palms it has been shown that the position of the axial triradius, *t*, is determined by heredity (Penrose 1954). In the case of finger-print patterns we know something of the inheritance of pattern-size. Patterns can be quantified, using the method of ridge-counting originated by Galton (1895) and developed by Bonnevie (1924). The number of ridges cutting or touching a straight line between the core of the pattern and the corresponding triradius are counted (see Fig. 3B). Where there are two triradii, two counts are made and the higher is used, while in the case of arches there is no count. The sum of the scores on all the fingers of an individual gives the *total ridge-count*. Values for total ridge-count vary between 0 and 300. The parent-child correlation coefficient for this measurement is 0.5 and the correlation between sibs is also very near a half (Holt 1952, 1956, 1957). Thus the size of finger-patterns is due to a number of perfectly additive genes. The correlation for total ridge-count between identical twins is very high, 0.95, while that between fraternal twins is of the same value as that between sibs. The effect of intra-uterine environment on finger-pattern size is small, amounting to about 5 per cent of the total variability. It appears, however, that environmental influences play a much greater part in determining palmar ridge arrangements.

Use of Finger-Prints in Twin Studies

Twin studies are useful for investigating various problems in biology and medicine. For most of these it is necessary to know with a minimum of error whether or not a twin pair is monozygotic. Dermatoglyphic features, particularly of fingers, are included among body traits widely used for diagnosis of zygotic type. Differences in total ridge-count between monozygotic twin pairs and between dizygotic twin pairs have been compared by various workers (e.g., Essen-Møller 1941, Geipel 1941, Holt 1952). All agreed in showing that about 80 per cent of monozygotic twins had differences of less than 20 ridges, compared with approximately 30 per cent of like-sexed dizygotic pairs. Further, no differences of more than 60 ridges were found among the monozygotic pairs, while in dizygotic pairs differences of up to 180 ridges were encountered. The fact that we now know something of the genetics of total ridge-count increases the usefulness of this measurement for diagnosing zygotic type. In recent years finger ridge-counts have been used in new ways for twin diagnosis. Two different approaches have been used: (1) by deciding the zygotic type on the basis of the relative probabilities of the hypotheses of the twins being monozygotic or dizygotic (Maynard, Smith and Penrose 1955, Lamy *et al.*, 1957); and (2) by the use of discriminant functions (Slater and Shields 1953, Nixon 1956). Methods such as these are useful in practice, particularly in conjunction with other diagnostic characters.

Palm-prints and Developmental Abnormalities

Since dermal ridge arrangements are in their complete and permanent form by the end of the fourth foetal month, they supply records of growth disturbances taking place in early pre-natal life. The best

example of this is found in mongolism, where there is retardation of growth of most parts of the body. The anomalies in the palm-prints and finger-prints of mongolian imbeciles will be described later.

In the case of zygodactyly of the third and fourth fingers, which is rarer than webbing of the second and third toes, Cummins and Midlo (1943) have shown that an interdigital triradius occurs on the palm near the site of the webbing. This interdigital triradius replaces the digital triradii *b* and *c*. Such an interdigital triradius may occur when no webbing of the fingers is evident. The presence of one is a minor expression of zygodactyly for 'interdigital triradii signify an intimate developmental relationship between neighbouring digits'.

Skeletal malformations of the hands and feet also affect ridge arrangement. In some cases of polydactyly there is an extra digital triradius on the palm, under the extra digit. It is thus sometimes possible to infer from a palm-print, even when scarring is absent, that a supernumerary digit has been amputated. Where a thumb is duplicated the two digits may each bear a finger pattern or the pattern on the first digit may be extended on the extra one. In the former event the patterns are usually of a similar type, but this is not always the case.

In extreme cases of skeletal deformities where there is reduction in number or fusion of the digits, such as ectrodactyly and acrocephalosyndactyly, there is much distortion of the dermatoglyphic patterns. In ectrodactyly, a rare inherited abnormality, the disturbance 'of the normal pattern of triradii is such that the ordinary points *a*, *b*, *c* and *d* on the palm are scarcely recognisable. When digits are missing the number of triradii is reduced and when there is syndactyly extra triradii are present'. (MacKenzie and Penrose 1951.)

Distortions in Dermal Ridge Arrangements Produced by Abnormal Chromosomes

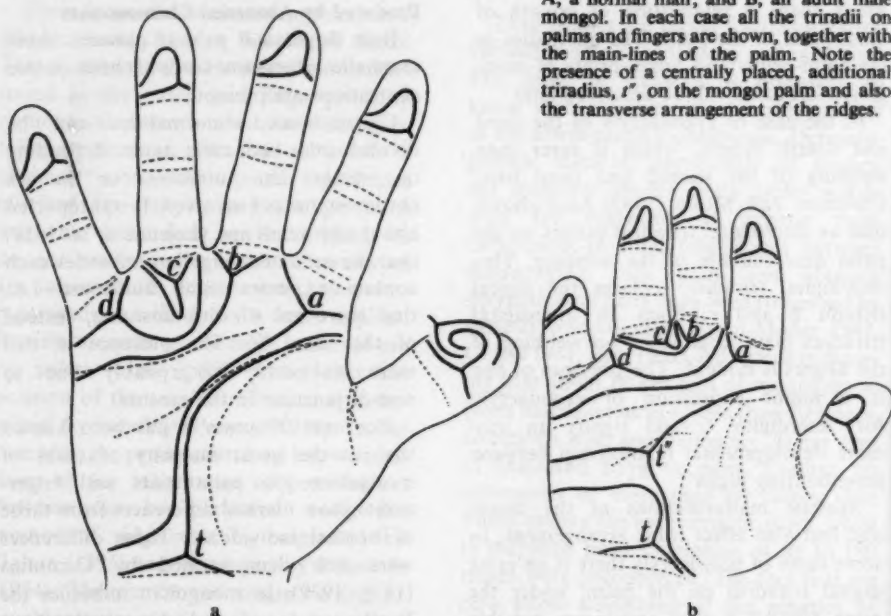
Both finger and palmar patterns show anomalies when some kinds of chromosome aberrations are present.

Chromosome abnormalities can be divided into two main types depending on whether the autosomes or the sex chromosomes are involved. It was reported about two years ago (Lejeune *et al.* 1959) that the cells of mongolian imbeciles each contain an extra small autosome—i.e., that there are 47 chromosomes, instead of the usual 46. The presence of this extra autosome is probably due to non-disjunction in the mother.

For over 20 years it has been known that in the great majority of cases of mongolism the palm-prints and finger-prints show marked differences from those of normal individuals. These differences were first demonstrated by Cummins (1936, 1939). In mongolian imbeciles the hands are broad and short with short, thick fingers. The palm is characterised by a transverse alignment of the dermal ridges and a high incidence in the hypothenar area of large patterns extending distally in the palm, associated with a triradius (*r'*) situated at or near the centre of the palm (see Fig. 5B). Thenar/first interdigital patterns are uncommon and when they occur are smaller and less complex than in the general population. Bimanual asymmetry is much reduced.

Finger-prints of mongols show less variation; there are fewer whorls and arches, the deficiency in both these patterns being compensated by an increase in the frequency of ulnar loops. The form of the loops tends to be high and almost L-shaped. Radial loops have a reduced frequency and are found chiefly on digits IV and V, especially on IV, instead of on digit II. The frequencies of the various pattern-types on the fingers of mongols (Cummins 1939) are as follows:

Fig. 5. Tracings of palm-prints of left hands of: A, a normal man; and B, an adult male mongol. In each case all the triradii on palms and fingers are shown, together with the main-lines of the palm. Note the presence of a centrally placed, additional triradius, t'' , on the mongol palm and also the transverse arrangement of the ridges.



Whorls	19.8%
Loops	{ Ulnar 75.4% } ..	77.6%
	{ Radial 2.2% }	
Arches	2.6%

Palm-prints are often of use in diagnosing mongolism at an early age. Cummins and Platou (1946) were able to classify 86 infants and children on palm-prints alone into three categories, mongols, normals and questionable. Agreement between palmar and clinical diagnoses was high—nearly 90 per cent. More recently, Ford Walker (1957) has described the use of dermal configurations in the diagnosis of mongolism.

Some dermatoglyphic traits of mongolism have been studied quantitatively. Penrose (1949, 1954) has used the *atd* angle to replace a qualitative differentiation of the triradii t and t'' . From palm-prints of families containing one or more mongol children he has shown that mongol children bear a greater resemblance to

their mothers than to their fathers, with respect to the position of the axial triradius. There is, therefore, some evidence in favour of the supposition that the genes affecting the position of t may be situated on the small autosome that is triplicated in mongolian imbeciles. The finger ridge-count has also been investigated and it has been shown that the mean finger counts in a sample of mongols differed from those in a control population (Holt 1951).

So far, less is known of distortions in dermal ridge arrangements associated with aberrations of sex chromosomes. In Turner's syndrome, however, where each cell contains only one X chromosome (i.e., a total of 45 chromosomes per cell), thenar/first interdigital patterns occur far more often than in the general population. Certain other anomalous features also appear to be characteristic of the condition.

In the one known case of Klinefelter-mongolism (Ford *et al.* 1959, Harnden



Plate 2A. Anterior view of the right hand in Klinefelter-mongolism. (From Harnden *et al.*, 1960).



Plate 2B. Palm-print of right hand of the Klinefelter-mongol.

et al. 1960) the cells were found to contain 48 chromosomes each, 45 autosomes + XXY—i.e., the extra small autosome found in other cases of mongolism and the extra X chromosome of Klinefelter's syndrome were both present. The dermal ridges on the palms were described as being characteristic of mongolism (Harnden *et al.* 1960). The anterior view of the hand is shown in Plate 2A, with a tracing of the triradii and main-lines of the palm in Fig. 6. A photograph of a palm-print is also shown (Plate 2B).

It has been seen that, in cases of abnormal development, ridge anomalies of two distinct kinds occur. Ridge configurations of the entire palm may be affected or the effect may be limited to a difference in frequency of a particular pattern as compared with general population.

Research on the dermatoglyphics of the palm and fingers has already produced interesting and significant results in the spheres of genetics and abnormal development. Investigations of this nature are in progress at the Galton Laboratory. Further research, particularly in the field of distortions produced by chromosome abnormalities, is likely to prove fruitful and may help to provide answers to many problems in medical biology.

Methods of Taking Palm- and Finger-prints

There are various ways of taking impressions of dermal ridges. Printer's ink is used for making dead-black impressions, but inkless methods offer various advantages. The principal methods of

Fig. 6. Triradii and main-lines of the dermal ridges as seen in Plate 2A. (From Harnden *et al.*, 1960).



printing are described by Cummins and Midlo (1943) in *Finger Prints, Palms and Soles*, Ch. 3, pp. 45-53. For a full description of the Faurot inkless method, with diagrams, see (Ford Walker 1957b).

It is important that palm-prints should be clear and complete. When taking finger-prints it is necessary to obtain rolled prints of the fingers, as well as plain impressions, to ensure complete prints of the patterns.

Acknowledgement: I am indebted to Prof. L. S. Penrose, F.R.S. for permission to reproduce Plates 2A and B, and for the palm-print of the Klinefelter-mongol.

SUMMARY

The uses of palm-prints and finger-prints in human biology are less well known than their uses for personal identification. So that the value of dermal ridge impressions in biology can be appreciated, a short account is given of the principal characteristics of ridged skin, together with a description of the main features of ridge arrangements on palms and fingers.

Attention is drawn to the use of dermal ridge prints in twin studies, and new ways of using finger-prints in diagnosing the zygotic types of twins are mentioned.

The value of palm-prints as records of growth disturbances in early foetal life is emphasised, anomalies in ridge arrangement in cases of zygodactyly, polydactyly, ectrodactyly and acrocephalosyndactyly being cited.

Distortions in dermal ridge configurations produced by aberrant chromosomes are described, the striking anomalies found on palm-prints of mongolian imbeciles being given in detail. It is suggested that this is an obvious and promising field for future research.

RÉSUMÉ

Empreintes de la paume et leur utilisation en biologie médicale

L'utilisation des empreintes de la paume et des doigts en biologie humaine est moins connue que sont utilisation pour l'identification personnelle. Afin que la valeur de la crête dermale puisse être appréciée en biologie, il est donné une brève description des principales caractéristiques de la peau striée ainsi que de la disposition des crêtes sur les paumes et sur les doigts.

L'auteur signale l'intérêt des empreintes des crêtes dermales dans l'étude des jumeaux et les nouvelles méthodes d'utiliser les empreintes digitales dans le diagnostic des types de jumeaux zygotes.

La valeur des empreintes de la paume pour suivre les troubles de croissance au début de la vie foetale est soulignée à la lumière d'exemples d'anomalies dans la disposition des crêtes dans des cas de zygodactylie, polydactylie, ectrodactylie et acrocéphalosyndactylie.

L'auteur décrit les distorsions de la configuration de la crête dermale dues à des chromosomes aberrants et présente en détail les anomalies frappantes qui apparaissent dans les empreintes de la paume chez les imbéciles mongoliens. C'est là, de l'avis de l'auteur, un champ évident et plein de promesses de recherches futures.

ZUSAMMENFASSUNG

Handflächenabdrücke und ihre Anwendung in medizinischer Biologie

Die Anwendung der Handflächen- und Finger-abdrücke in der menschlichen Biologie ist weniger bekannt, als die Anwendung für Identifizierung der Personen. Um den Wert des Abdruckes des Lederhautkammes in Biologie abschätzen zu können, werden die hauptsächlichsten Kennzeichen der gefurchten Haut sowie die Anordnung der Kämme auf den Handflächen und Fingern kurz beschrieben.

Die Bedeutung der Abdrücke der Hautkämme in der Zwillingsforschung und neue Methoden für die Anwendung der Fingerabdrücke für die Diagnose der zygotischen Typen der Zwillinge werden erwähnt.

Der Wert der Handflächenabdrücke für die Forschung des foetalen Lebens wird betont mittels Anführung von Anomalien der Anordnung der Kämme bei Zygodaktylie, Polydaktylie, Ektrodaktylie und Akrokephalosyndaktylie.

Verzerrungen der Gestalt der Hautkämme, erzeugt durch abweichende Chromosome, werden beschrieben und die bei monogoloiden Imbezillen gefundenen auffallenden Abnormitäten der Handflächenabdrücke werden ausführlich dargestellt. Der Autor ist der Meinung, dass es sich hier um ein augenscheinliches und für künftige Forschungen vielversprechendes Gebiet handelt.

REFERENCES

- Bonnevie, K. (1924) 'Studies on papillary patterns of human fingers.' *J. Genet.* **15**, 1-112.
- Cummins, H. (1936) 'Dermatoglyphic stigmata in mongolian idiocy.' (Abstract) *Anat. Rec.* **64**, (Suppl. 2), 11.
- (1939) 'Dermatoglyphic stigmata in mongoloid imbeciles.' *Ibid.*, **73**, 407-15.
- Midlo, C. (1926) 'Palmar and plantar epidermal ridge configurations (dermatoglyphics) in European-Americans.' *Amer. J. Phys. Anthropol.*, **9**, 471-502.
- , —, (1943) *Finger Prints, Palms and Soles*. Philadelphia: Blakiston Company.
- , Platon, R. V. (1946) 'Mongolism: an objective early sign.' *Sth. med. J.* **39**, 925-8.
- Essen-Moller, E. (1941) 'Empirische Ähnlichkeitsdiagnose bei Zwillingen.' *Hereditas*, **27**, 1-50.
- Ford, C. E., Jones, K. W., Miller, O. J., Mittwoch, U., Penrose, L. S., Ridler, M., Shapiro, A. (1959) 'The chromosomes in a patient showing both mongolism and the Klinefelter syndrome.' *Lancet*, **i**, 709.
- Ford Walker, N. (1957a) 'The use of dermal configurations in the diagnosis of mongolism.' *J. Ped. St. Louis*, **50**, 19-26.
- (1957b) 'Inkless methods of finger, palm and sole printing.' *Ibid.*, 27-29.
- Galton, F. (1892) *Finger Prints*. London: Macmillan.
- (1895) *Finger-print Directories*. London: Macmillan.
- Geipel, G. (1941) 'Die Gesamtanzahl der Fingerleisten als neues Merkmal zur Zwillingsdiagnose.' *Z. Morph. Anthr.* **39**, 414-9.
- Harnden, D. G., Miller, O. J., Penrose, L. S. (1960) 'The Klinefelter-mongolism type of double aneuploidy.' *Ann. hum. Genet. Lond.*, **24**, 165-169.
- Holt, S. B. (1951) 'A comparative quantitative study of the finger-prints of mongolian imbeciles and normal individuals.' *Ann. Eugen., Lond.*, **15**, 355-74.
- (1952) 'Genetics of dermal ridges: inheritance of total finger-ridge count.' *Ibid.*, **17**, 140-61.
- (1956) 'Genetics of dermal ridges: parent-child correlations for total finger ridge-count.' *Ann. hum. Genet.*, **20**, 270-81.
- (1957) 'Genetics of dermal ridges: sib pair correlations for total finger ridge-count.' *Ibid.*, **21**, 352-62.
- Lamy, M., Frézal, J., de Grouchy, J., Kelley, J. (1957) 'Le nombre de dermatoglyphes dans un échantillon de jumeaux.' *Ibid.*, **21**, 374-96.
- Lejeune, J., Gautier, M., Turpin, R. (1959) 'Les chromosomes humains en culture de tissus.' *C.R. Acad. Sci., Paris*, **248**, 602-3.
- MacKenzie, H. J., Penrose, L. S. (1951) 'Two pedigrees of ectrodactyly.' *Ann. Eugen., Lond.*, **16**, 88-96.
- Maynard Smith, S., Penrose, L. S. (1955) 'Monozygotic and dizygotic twin diagnosis.' *Ann. hum. Genet.*, **19**, 273-89.
- Nixon, W. L. B. (1956) 'On the diagnosis of twin-pair ovularity and the use of dermatoglyphic data.' *Novant' anni delle Leggi Mendeliana*, pp. 235-45. (Ed. L. Gedda). Rome: Istituto Gregorio Mendel.
- Penrose, L. S. (1949) 'Familial studies on palmar patterns in relation to mongolism.' *Proc. 8th int. Cong. Genet. (Hereditas, suppl. vol.)* 412-6.
- (1954) 'The distal triradius *t* on the hands of parents and sibs of mongol imbeciles.' *Ann. hum. Genet.*, **19**, 10-38.
- Slater, E., Shields, J. (1953) 'Psychotic and Neurotic Illnesses in Twins. *Spec. Rep. Ser. med. Reg. Com., Lond.*, no. 278.

NOTICE

The Bristol Royal Hospital for Sick Children

THE hospital illustrated on the cover of this *Bulletin* was founded by Mark Whitwill, its first president and treasurer, who gave this account of the origin of the hospital, then known as *The Hospital for Sick Children and for the Outdoor Treatment of Women*, in his annual report for 1891:

'The Out-Patients work, which is now so important a feature, was begun in the year 1857 by Dr. Mortimer Granville, then a Surgeon in Bristol, who opened a room in Lower Castle Street, and having, in conjunction with Mr. W. Ormerod, interested some gentlemen in the matter, a Committee was formed, a Staff organised, and the Institution transferred to a house in St. James's Square. During the early years it had a hard struggle for life. In 1864 your President was invited to join that Committee, and he suggested that they should not be content with a Dispensary merely, but that an attempt should be made to found a Hospital.'

Mr. Whitwill was a man with ideas in advance of his time. According to the first annual report, for 1867, the objects of the institution were 'to provide for the reception, maintenance, medical and surgical treatment of children under twelve years of age in a suitable building cheerfully and salubriously placed . . ., to promote the advancement of medical science . . . and to provide for the instruction of students in these essential departments of medical knowledge'. And again, 'to diffuse among all classes of the community, and particularly among the poor, a better acquaintance with the management of infants and children during *health* and sickness'.

By the First World War the work of the hospital had much increased and there was an average of 900 children in the wards each year and also about 100 women. The admission of women as inpatients did not cease until the hospital was bombed in the Second World War. The 89 children in the hospital at the time were all safely evacuated, but the building was so badly damaged that it could not be used for three months and the out-patient department was completely destroyed. Rebuilding and extension have therefore been a major activity since the war. In 1948 the hospital ceased to be a voluntary body and became part of the new teaching hospital designated in the National Health Act.

The Royal Hospital for Sick Children had had students walking the wards since the 1890's; but the unity established by the Act has led to a closer affinity between the new Board of Governors, the Medical School and other associated bodies, especially the local authorities and the statutory health service. Research work has always been an activity of the hospital and is gradually increasing, close co-operation being maintained with other University departments. Long-term research is being carried out on the diagnosis, treatment and education of children with cerebral palsy; the causes and consequences of retardation are under special clinical and biochemical investigation. There is an advisory service to parents connected with the study of clinical genetics.

This September, the National Spastics Society is organising a conference in Bristol on the management of hemiplegic cerebral palsy. Those attending it will be looking forward to visiting the Bristol Royal Hospital for Sick Children and seeing for themselves some of the features of this famous children's hospital.

Spasticity and Spasms in Hemiplegia and Paraplegia*

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SPASTICITY is a state of excessive neuromuscular activity characterised clinically by induced progressive increased resistance or tension of muscles during stretch, after a variable free interval, and is usually associated with lengthening or 'clasp-knife' reactions, hyperactive deep tendon reflexes and clonus. Often there are other phenomena present, such as paralysis of willed movement and abnormal reflexes such as the Babinski. Spasms of both flexor and extensor muscles in the affected extremities may appear in addition to spasticity, as a different type of muscle response. These conditions are the result of lesions in the central nervous system involving so-called upper motor neurones or corticospinal pathways and their interconnections at all levels. They usually, therefore, occur in patients with central nervous system disease which has produced hemiplegia and paraplegia.

Mechanism of Spasticity

That the spastic state is basically due to exaggeration of monosynaptic spinal stretch reflexes is one of the great contributions of Sherrington and his group (Sherrington 1906, Liddell and Sherrington 1924). Their studies defined the nature of the stretch reflex in both the decerebrate and spinal animal, and emphasised the extensor or tonic predominance of reflex

activity in the decerebrate and the flexor or phasic activity in the spinal.

Walshe (1919), applying these principles to lesions in man, presented the following argument: in spastic paralysis of the lower limbs there are two clinical types, the extended and the flexed; in a hemiplegia due to a capsular lesion the extended type is constant, irrespective of the degree of pyramidal involvement. In a typical hemiplegia, the lower limb lies fully extended with the foot plantar-flexed; the limb extensors are hypertonic and have brisk tendon reflexes and often clonus. These extensor muscles also retain relatively more voluntary power than their flexor antagonists. The flexors, on the other hand, do not show the spasticity characteristic of the extensors, although they yield brisk tendon jerks and show a phasic type of reflex activity of which the flexion reflex and its component Babinski response are the chief expressions. In severe bilateral spinal lesions, the flexed type of spastic paraplegia predominates. The extensors lose their characteristic reflex activity, while their flexor antagonists show an extreme degree of reflex action producing a flexed position of the limbs and flexor spasms. The two reflex systems, tonic and phasic, initially described by Sherrington, could be, therefore, differently and separately affected.

Walshe also concluded that the extended type of spastic paralysis was identical with

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the decerebrate rigidity described by Sherrington in animals, and that the spastic element was the same phenomenon as the tonic extensor rigidity seen in these preparations, possessing a similar mesencephalo-spinal reflex arc, whereas the phasic reflex activity of the flexors seen in both types of paralysis was spinal in level and was the expression of the reflex activity of the spinal cord. The flexion attitude of the upper limb in hemiplegia, however, was traced to the altered functions of the upper limb in man and its lack of activity in locomotion.

Brain (1927) demonstrated that bending a patient with hemiplegia forwards, to assume a quadrupedal posture, produced an extensor attitude in the spastic upper extremity. Extensor tonus is therefore ascribed to the proprioceptive mechanisms concerned mainly with posture. Denny-Brown (1950) emphasised that the greater the extent of proprioceptive spinal network that is isolated from the brain-stem, the more completely is spasticity developed, disproportionately releasing antigravity muscles of both upper and lower extremities from higher inhibitory influences.

Clinical Evaluation

Clinical evaluation of patients with spastic hemiplegia, characteristically secondary to subcortical cerebral infarction, reveals the following: progressive resistance to stretch after a variable free interval, a preponderance of flexor activity in the upper extremities at the elbow, wrist and fingers, and in the lower extremities a preponderance of extensor activity at the knee and ankle, with plantar flexion. Adductor spasticity of the thigh is often severe. There is hyperreflexia of the deep tendon reflexes especially at the knee, biceps and ankle. The abnormally released reflexes in the Babinski group are present in the affected extremities and often do not correlate in intensity with any of the

other signs, such as the degree of spasticity or hyperreflexia. In evaluating voluntary motor function, there is often no distinct correlation between intensity of spasticity and residual motor activity. As is well known, lesions involving precentral motor cortex and pyramidal pathways may produce paralysis and hyperreflexia with only minimal or no spasticity. In children the intensity of hemiplegic spasticity may be altered by such mechanisms as the tonic neck reflexes. Twitchell (1951) has emphasised that the stage of spastic dominance with little independent willed movement in capsular hemiplegia is one of proprioceptive hyperactivity. In recovering hemiplegia, spasticity diminishes as the instinctual grasp reflex returns.

Spasticity in individuals with spinal paraplegia may show either flexor or extensor predominance, although usually the former. Extensor predominance is usually associated with incomplete transecting lesions of the spinal cord, but even in these instances a shift to flexor preponderance may occur. Also, extensor spasticity has been found in patients with complete transecting lesions. Lengthening or clasp-knife responses usually occur in the paraplegic patient with extensor spasticity as in most hemiplegics. The deep reflexes in spastic paraplegia are usually hyperactive at the ankle, with clonus. The hamstring or flexor reflexes are active in patients with predominantly flexor spasticity; the knee-jerk is elicited only inconsistently in patients with extensor predominance. Again, the abnormal reflexes, such as the Babinski, are usually present in the extremities involved, but do not correlate in intensity with the other signs. Adductor spasticity is often prominent along with flexor paraplegia.

Investigations applying electromyography of the spastic stretch reflex in man (Hoefler and Putnam 1940) have shown electrical silence of the muscles at rest, a lower

reflex threshold together with enlarged reflexogenic area, augmented response, a tendency towards repetition producing clonus, synchronisation in agonist and antagonist muscles, and spread or irradiation.

Spasms in Hemiplegics and Paraplegics

Spasms, or rapid involuntary muscular contractions, occur in spastic muscles in these states as separate phenomena, appearing seemingly spontaneously or induced by a variety of overt stimuli. These reactions may appear in patients with either spinal or supraspinal spasticity. They may be quite painful and present a serious therapeutic problem. Spasms experienced by hemiplegic patients are most often extensor in the affected lower extremity, and are frequently associated with spontaneous clonus at the ankle. Infrequently, flexor spasms of the upper extremity occur in the hemiplegic. Some patients experience spasms of the abdominal muscles. Spasms may appear many times daily and may last several minutes. Stretching of the muscles of the lower extremity may act as a stimulus to spasm as well as spasticity. However, a frequent stimulus in the hemiplegic, occurring in many patients, is merely lying quietly in bed awaiting sleep. Other stimuli include bladder distention, bathing or stroking the skin.

Spasms in spinal paraplegic patients are more common, usually more severe and painful, and often induce bladder emptying. Patients with predominantly extensor activity in the affected limbs usually experience extension spasm, but occasionally even flexion may occur. Patients with predominantly flexor spasticity in the lower extremities mainly experience flexion spasms, but occasionally an extension occurs spontaneously. In most patients, either hemiplegic or paraplegic, extensor spasms involve thigh adductors and inver-

tors of the foot in addition to the major muscles. The usual stimuli in the paraplegic are bladder distention, passive stretching, skin stroking, uncomfortable positioning of the limbs, or often merely a change of posture in bed. The spinal paraplegic patients also experience spasms while lying quietly in bed before sleep. Spasms in all patients are more severe and frequent during fatigue.

Mechanism of Exaggerated Responses

The basic causative factors involved in these exaggerated reflex responses have been the subject of many investigations, but complete clarification has still to be attained. There appears to be a dualistic mechanism (Magoun and Rhines 1947): the impairment of central inhibitory influences on the stretch reflex arc with a release effect, and a maintained or probably exaggerated activity of central facilitatory influences, which may augment spinal stretch reflexes. Although spasticity is usually associated with lesions of the so-called pyramidal corticospinal system, the non-pyramidal motor projection systems or extrapyramidal pathways also are implicated. In recent years, the mesodiencephalic brain-stem reticular formation, in its inhibitory and facilitatory functions, has been emphasised as a major central mechanism of spinal motor neurone control (Magoun 1958, Ward 1958). These supraspinal systems are not homogeneous and undifferentiated, but there are specific nuclei and fibre tracts with definite facilitatory or inhibitory effects on the spinal neurones. The integrity of the brain-stem complex up to and including the Deiter's nucleus of the vestibular complex appears to be necessary for the production of extensor spasticity. The function of the anterior lobe of the cerebellum also is related to the development of this type of spasticity. A profound influence of the fastigial nucleus has been

demonstrated by Moruzzi and Pompeiano (1956). Damage to the red nucleus and its connections also may be involved in loss of righting reflexes and exaggeration of antigravity postures.

The stretch reflexes are dependent on proprioceptive impulses derived from the affected muscles, tendons and joints. Denny-Brown (1950, 1960) has emphasised the susceptibility of these reflexes to proprioceptive stimuli. The abolition of contact reactions in hemiplegia aids in the release of the proprioceptive stretch reflexes. In recent years there has been a re-evaluation of these mechanisms in relation to muscle spindle control by gamma efferent system activity (Granit 1955). The muscle spindles are in parallel with the extrafusal or motor fibres, and so situated that the sense organs within the spindles become unloaded in silence when the alpha motor fibres are made to contract, especially if the contraction is a response to stimulation. The afferents from the muscle spindles make contact with the ventral horn cells and thus contribute to the monosynaptic reflex. The alpha ventral horn cells giving rise to ordinary motor fibres can be driven indirectly through the gamma efferents to the spindles, as well as directly by other converging motor pathways. Gamma activity is considered to be truly tonic and persistent except when actively inhibited. The muscle spindles are similarly kept discharging in tonic fashion and thus facilitate discharge of the ventral horn cells. However, adaptation occurs in the muscle spindle receptor system and is an important factor in the reflex control system over muscle length and tension (Partridge and Glaser 1960, Matthews 1959). Further, Granit (1956) has demonstrated spindle stretch reflex rebound by post-tetanic potentiation and temporal summation, and has correlated this with spasticity. Single ventral horn alpha cells

are regarded to have been made 'spastic' by this mechanism. Thus, tonic gamma effects contribute to setting the level of excitability in the spinal cord by enhancing the activity in the muscle spindle receptor organs (Hunt and Perl 1960). The supraspinal control over the gamma system, both for initiating and inhibiting activity, lies in the reticular formation and in associated cerebellar centres.

In decerebrate rigidity of the Sherrington type, the spastic animal has exaggerated stretch reflexes with very high tonic gamma activity (Granit 1955, Matthews and Rushworth 1957, Glaser and Partridge 1958). This type of spasticity, therefore, may be due to hyperactivity in the gamma system maintaining excessive excitation of the ventral horn cells by the spindle loop. However, in animals with anterior cerebellar damage in addition to decerebration, there is marked spasticity with 'passive' behaviour of the gamma system, and maintenance of spasticity despite dorsal root section or selective removal of gamma activity by peripheral-nerve or muscle anaesthesia with procaine (Granit 1955, Matthews and Rushworth 1957, Glaser and Partridge 1958). This has been called the alpha type of spasticity. On the contrary, the gamma type of spasticity in the Sherringtonian decerebrate animal can be melted either by selective anaesthetic block of gamma components of peripheral nerve or by injecting procaine into the muscles involved (Matthews and Rushworth 1957).

These different types of spasticity probably have their clinical counterparts in human subjects, depending on the intensity of cerebellar system involvement and capable of being analysed by selective pharmacological block (Rushworth 1960). However, in human cases of spasticity, although these blocking procedures may relieve the spasticity (Landau *et al.* 1960, Rushworth 1960) there does not appear to

be any clear indication of excessive gamma discharge (Landau *et al. loc., cit.*). Severe spasms and spasticity in paraplegia have been relieved by the intrathecal instillation of phenol, to destroy anterior roots (Nathan 1959). The preservation and even improvement of some voluntary motor activity in the absence of proprioceptive reflexes suggests a selective effect with mainly gamma efferent destruction in these cases, although in others both alpha and gamma efferents are destroyed, with more flaccid paresis ensuing.

Other Possible Mechanisms

The usual intense flexor spasticity of paraplegia and sensitivity to flexor spasms may be related to increased or released reflex activity secondary to lowering of threshold of spinal neurones. However, the differences between the phenomena from spinal and supraspinal lesions require the consideration of other possible mechanisms. The special tendency to flexor spasms may be related to scarring, particularly in traumatic cases, at the distal stump of the spinal cord, setting up irritative impulses in afferent pathways having reflex collaterals in the affected segments (Scarff and Pool 1946). Another probably more significant factor may be related to changes at the innervation of the interneurone. Normally, the surface of the interneurone is saturated with boutons. When those from descending tracts degenerate, the remaining terminals, chiefly of afferent fibres, may sprout new boutons (McCouch *et al.* 1958). After cord transection, potentials at the interneurone attributed to afferent terminals are significantly increased. This excessive activity might be produced by the sprouting and be responsible for spasticity (McCouch *et*

al. loc., cit.). Section of the appropriate dorsal roots vacates bouton surface sites on interneurons, tends to eliminate the tendon reflexes and may relieve spasticity, but only partially. Exaggeration of stretch reflexes, therefore, may not necessarily be the sole cause of spasticity or spasms after spinal injury. The evidence, however, does not suggest that the threshold of interneurons or of motor neurones is lowered below normal levels.

It has been shown that ischaemic lesions of the lumbar spinal cord in animals, causing loss of cells in the intermediate grey matter, produce continued spastic responses and spasms, even when the cord is transected above the lesion (Gelfan and Tarlov 1959). The spasticity in these instances is thought to be due to the loss of the inhibitory effect of the small interneurons, particularly of the Renshaw type, on the anterior horn cells (Hunt and Perl 1960). The spasms may persist even in the absence of all afferents and supraspinal connections. It may be concluded that complete deprivation of all internuncial neurones results in uncontrolled spontaneous discharge of motor neurones together with the increases in monosynaptic reflex activity. Spasticity, and its accompanying release or increase of stretch reflex activity, may, therefore, be associated either with loss of control over interneurons or with actual loss of interneurons. The presence of collaterals in various segments determines the distribution of the responses in the spastic extremity. In cervical spinal cord lesions at or about the fifth cervical level, for example, severe spasms may occur in addition to spasticity, with abduction of the arms and flexion of the forearms (Penry *et al.* 1960), producing an unusual forced posturing.

SUMMARY

There are differences between the spasticities appearing in hemiplegia due to supraspinal lesions and in paraplegia due to spinal lesions, although the basic quality is progressive increased tension in muscles in response to stretch. Spasticity in hemiplegia is characterised by extensor predominance in the lower extremity and flexor predominance in the upper. In spinal paraplegia the spastic lower extremities are usually in flexion, although occasionally extension is present, more so with incomplete spinal lesions. Spasms, or rapid involuntary muscular contractions, have a similar distribution in the two states and may be spontaneous or induced by a variety of stimuli.

The general physiological mechanisms involve exaggerated spinal stretch reflexes, due to release from and facilitation by supraspinal controls, especially reticular and cerebellar influences, increased susceptibility to proprioceptive stimuli, and possible overactivity in the gamma control system of the muscle spindles.

Other special factors in spinal lesions include irritative phenomena at the site of the lesion and excessive activity at interneurons related to sprouting of boutons.

REFERENCES

- Brain, W. R. (1927) 'On the significance of the flexor posture of the upper limb in hemiplegia, with an account of a quadrupedal extensor reflex.' *Brain*, **50**, 113-137.
- Denny-Brown, D. (1950) 'Disintegration of motor function resulting from cerebral lesions.' *J. nerv. ment. Dis.*, **112**, 1-45.
- (1960) 'Motor mechanisms—introduction: the general principle of motor integration.' *Handbook of Physiology. Neurophysiology II. Chapter 32*, pp. 781-796.
- Gelfan, S., Tarlov, I. M. (1959) 'Interneurons and rigidity of spinal origin.' *J. Physiol.*, **146**, 594-617.
- Glaser, G. H., Partridge, L. D. (1958) 'An experimental study of spasticity.' *Trans. Amer. neurol. Ass.*, **83**, 159-160.
- Granit, R. (1955) *Receptors and Sensory Perception*. New Haven, Conn.: Yale Univ. Press.
- (1956) 'Reflex rebound by post-tetanic potentiation. Temporal summation-spasticity.' *J. Physiol.*, **131**, 32-51.
- Hoefner, P. F. A., Putnam, T. J. (1940) 'Action potentials of muscles in "spastic" conditions.' *Arch. Neurol. Psychiat.*, **43**, 1-21.
- Hunt, C. C., Perl, E. R. (1960) 'Spinal reflex mechanisms concerned with skeletal muscle.' *Physiol. Rev.*, **40**, 538-579.
- Landau, W. M., Weaver, R. A., Hornbein, T. F. (1960) 'Fusimotor nerve function in man. Differential nerve block studies in normal subjects in spasticity and rigidity.' *Arch. Neurol.*, **3**, 10-23.
- Liddell, E. G. T., Sherrington, C. S. (1924) 'Reflexes in response to stretch (myotatic reflexes).' *Proc. Roy. Soc.*, **96B**, 212-242.
- McCouch, G. P., Austin, G. M., Liu, C. N., Liu, C. Y. (1958) 'Sprouting as a cause of spasticity.' *J. Neurophysiol.*, **21**, 205-216.
- Magoun, H. W. (1958) *The Waking Brain*. Springfield, Ill.: C. C. Thomas, pp. 15-32.
- Rhines, R. (1947) *Spasticity. The Stretch Reflex and Extrapyramidal Systems*. Springfield, Ill.: C. C. Thomas.
- Matthews, P. B. C. (1959) 'A study of certain factors influencing the stretch reflex of the decerebrate cat.' *J. Physiol.*, **147**, 547-564.
- Rushworth, G. (1957) 'The selective effect of procaine on the stretch reflex and tendon jerk of soleus muscle when applied to its nerve.' *Ibid.*, **135**, 245-262.
- Moruzzi, G., Pompeiano, O. (1956) 'Crossed fastigial influence on decerebrate rigidity.' *J. comp. Neurol.*, **106**, 371-392.
- Nathan, P. W. (1959) 'Intrathecal phenol to relieve spasticity in paraplegia.' *Lancet* **ii**, 1099-1101.
- Partridge, L. D., Glaser, G. H. (1960) 'Adaptation in regulation of movement and posture. A study of stretch responses in spastic animals.' *J. Neurophysiol.*, **23**, 257-268.
- Penry, J. K., Hoefnagel, D., Van Den Noort, S., Denny-Brown, D. (1960) 'Muscle spasm and abnormal postures resulting from damage to interneurons in spinal cord.' *Arch. Neurol.*, **3**, 500-512.
- Rushworth, G. (1960) 'Spasticity and rigidity: An experimental study and review.' *J. Neurol. Neurosurg. Psychiat.*, **23**, 99-118.
- Scarff, J. E., Pool, J. L. (1946) 'Factors causing massive spasm following transection of the cord in man.' *J. Neurosurg.*, **3**, 285-293.
- Sherrington, C. S. (1906) *Integrative Action of the Nervous System*. New Haven, Conn.: Yale Univ. Press.
- Twitchell, T. E. (1951) 'The restoration of motor function following hemiplegia in man.' *Brain*, **74**, 443-480.
- Walshe, F. M. R. (1919) 'On the genesis and physiological significance of spasticity and other disorders of motor innervation: With a consideration of the functional relationships of the pyramidal system.' *Brain*, **42**, 1-28.
- Ward, A. A. (1958) 'Efferent functions of the reticular formation.' In: *Reticular Formation of the Brain*. Boston: Little, Brown and Co. Pp. 263-273.

Vocational Training for the Cerebral Palsied

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EXPERIENCE gained in the National Spastics Society's first Vocational Training Centre has shown that a large proportion of cerebral palsied adults are capable of earning their own living in open employment. Employers are understandably shy of accepting responsibility for training handicapped persons of a class hitherto regarded as unemployable. The centre exists to give them that training in ordinary industrial circumstances.

The National Spastics Society's training centre at Sherrards was conceived in 1954 when few people with cerebral palsy could hope to earn their living in open industry. Some other handicapped people, such as the blind, qualified automatically for the benefits of welfare services provided by statute in virtue of their handicap, but these 'spastics' were not understood. They were largely shunned by the authorities responsible for the welfare of the physically handicapped. When Sherrards was opened in 1956, there was no other residential centre in the world exclusively for training the cerebral palsied, and those charged with its development therefore had no experience of others to guide them. No statistics were available to indicate the demand for such an institution but the need was obvious to the National Spastics Society, both for training known 'spastics' of post-school age and for giving vocational training to some of those who were to be educated in the Society's own schools. Apart from obtaining suitable premises, three basic problems emerged: (1) the

selection of candidates; (2) the scope of training to be given; and (3) the placement of the trainees in open employment.

Selection of Candidates

Qualifications and Nominations.

It was decided to open Sherrards for cerebral palsy patients of either sex, in the 16-25-year age-group, who according to our concept were capable of successful training. Applications were made on the patients' own initiative or through agencies such as the Ministry of Labour, hospital almoners, local authority education and welfare departments, and the Society's own supporters and workers.

Interview Technique.

Of the 236 applicants interviewed so far, the records of 25 are no longer available, but those of 211 have been analysed. Each was interviewed and examined for about an hour by the medical consultant, followed by about half an hour with a lay panel headed by the Principal of the centre. At least one parent was expected to be present throughout. The interviews were followed by a conference, when the findings at each examination were discussed and a joint recommendation for admission or rejection was made.

The medical examination, not over-riding except in respect of definite medical reasons for rejection, was conducted to confirm the diagnosis of cerebral palsy and exclude cases in which the Society has no concern. Such cases included inherited,

acquired, and progressive forms of neurological or psychological disorder. Apart from disqualifications, the examination sought to reveal the applicant's physical competence and functional capacity, rather than his incompetence and incapacity, and to evaluate the background features of upbringing, education and social and domestic status. Conventional intelligence tests were not applied and a formal intelligence quotient was not calculated. After an hour and a half's contact during the interviews, when total demeanour was studied critically and continuously, intelligence was graded as better than average, average, low or very low.

Analysis of Applications.

The 211 applicants whose case-papers have been studied fell into the following categories of locomotor disorder:

Left hemiplegia	26
Right hemiplegia	16
Double hemiplegia	10
Monoplegia	8
Quadriplegia	48
Paraplegia	44
Diplegia	3
Athetosis	34
Others	22

Nearly 50 per cent of the cerebral palsied have a dual handicap. These were as follows:

Severe dysarthria	49
Mutism	3
Severe deafness	10
Blindness	1
Congenital cataracts	1
Gross nystagmus	2
Hemianopia	2
Epilepsy	23

Of the 136 males, 90 were recommended for admission and 46 for rejection. Of the 75 females 54 were suitable and 21 unsuitable. The 67 unsuccessful candidates were rejected for the following reasons:

Motor handicap not permitting any hand function, attention to personal toilet, or travelling to and from work after training	26
Intelligence not sufficient for comprehension of simple instructions	22

Blindness	1
Major epilepsy	12
Belligerent temperament	1
Other diseases	3
Affliction slight enough to warrant direct placement in industry	2

Scope of Training Facilities.

Sherrards is a country house standing in some 7 acres of land. The present residential accommodation is sufficient for only 31 cases, but more workshops have been built and a new hostel is under construction which will increase the accommodation to 60 places. A ramp has replaced a front doorstep and each staircase has been fitted with a second hand-rail. Beyond that, the furnishings and equipment are all standard, without special modifications or adaptations. Under the direction of the Principal, a house-keeper-matron superintends the work of male and female house parents and the domestic staff. Under the same direction a senior instructor manages the workshops and controls a staff of 4 assistant instructors. General medical attention is provided by a local practitioner, and the specialist problems in physical medicine are dealt with by the medical consultant, assisted by a physiotherapist and speech therapist. A teacher is provided to give lessons in reading, writing and arithmetic on 2 evenings a week, and a class in ball-room dancing is held once a week, under the local authority further education scheme. Guest speakers lead discussions or lecture on current affairs once a week and ministers of religion are invited to conduct services on Sundays.

Training.

Training is given in engineering, wood-working, and printing. A domestic science block is being built and training will be offered in this subject and in commerce. However, it should be pointed out that 'socialisation' has been found to be an

essential function of the centre and to rank equally in importance and difficulty with technical instruction.

Technical Equipment.

Engineering:

Lathes—capstan and centre.
Drills—pillar and bench.
Milling—horizontal.
Presses—fly, kick and toggle.
Welding—spot welder.
Injection moulders.
Grinders—bench and drill grinders.
Guillotine.
Folding machine.
Shears—bench hand type.
Bench hand tools—files, hacksaws, drills, rules and marking tools, micrometer gauges, screwdrivers, spanners and hammers.

Woodworking:

Mortising machine—pillar.
Lathe—woodturning.
Saws—fixed and tilting table.
Planers.
Thicknessers.
Sanders—disc and belt.
Bench hand tools—saws, chisels, hammers, screwdrivers, planes, braces, squares and rulers and scribers.

Printing:

Adana hand-operated printing machine and flat-bed.
Auto numbering machine.
Founts of type and type cases.

Miscellaneous:

Time clock.
Dust extractor.

Placement in Employment

When this article was drafted (August 1960), 87 cases had been admitted out of 144 on the waiting-list; 31 were still in residence, 34 placed in open employment and 23 discharged.

Job placements were as follows:

Press operators	2
Capstan lathe operators	1
Tickopress operator	3
Bench assemblers	10
Packers	2
Inspectors	3
Box makers	2
Labourer	1
Usherette	1
General assistant	1
General machinist	1
Spot welder	2
French polisher	1
Fitter	2
Copytypist	1
Printer	1

The average period of residence in the centre was 10 months. The shortest period was 2 months and the longest, 27 months. Of the 34 trainees placed in employment, 30 are still employed; 1 was discharged as redundant; 1 gave up his job; and 2 lost their positions from inefficiency.

Since this article was drafted a further 7 cases have become employed:

Capstan lathe operator	3
Labourer	1
Clerk	1
Driller	1
Assembler	1

In this group the average training period was 19 months, the shortest being 9 and the longest 34 months.

Discharges were for the following reasons:

Untrainable	8
Misbehaviour in hostel	3
Laziness	1
Homesickness	1
Intercurrent illness	1
Incontinence	1
Psychopathic behaviour	2
Epilepsy	1
Resignation	2
Withdrawal by parents	2

The Centre in Operation

Sherrards is recognised as a sheltered workshop, and in order to qualify for grants towards training and maintenance, each entrant is registered as a disabled person. After a period of training, during which representatives of the Ministry of Labour and the Principal hold frequent case conferences, the trainee reaches a state of proficiency at work which qualifies him or her for the status of temporary employee. The personal grant then ceases, but thereafter each receives a wage paid weekly in cash, under the Truck Acts, by the employer—the National Spastics Society. A charge is made by the Society for residential accommodation which is

deducted from the wage and thus each temporary employee learns something of self-support and independence.

In the Workshops.

Each trainee is allotted frequently changing tasks with different neighbours for about a fortnight after entry. This is a period of assessment of physical and functional capacity, of personality, temperament and intelligence. Then, having regard to these factors and to the prospects of employment in the trainee's own home area, they are drafted to one of the workshops. The procedure is not rigid and each assessment is constantly reviewed. This is necessary because a completely new training problem may present itself to the Principal and Senior Instructor as each case of cerebral palsy responds to the total treatment. Social life and education classes within the hostel may enhance the trainee's attitude and mental ability to enable him to do semi-skilled work requiring thought, judgment or measurement, and purpose-planned surgical procedures, carried out as a result of appraisal, may radically improve his dexterity or mobility.

Every task performed in the workshops is a constructive, productive job, identical with that carried out in open industry by non-handicapped persons. The Principal seeks work on a sub-contracting basis at the best price he can negotiate. The worker has to realise the importance of efficiency, craftsmanship and productivity and this objective is clouded if he knows his efforts are subsidised. Every effort is therefore made to make the workshops self-supporting. As examples, contracts have been secured for punching components of tabulating machines, manufacturing bits for soldering irons, constructing insulated electrode holders for arc welding, making small collecting boxes and large packing cases in timber, and printing pamphlets, stationery and tickets. Practical experience

is enlarged by the variety of these tasks, and, since many processes are involved in producing the finished article, the jobs can be broken down to permit allocation of work to those who can fetch and carry, measure and mark off, operate a machine, finish, or assemble and pack. At all stages the instructors give instruction to individuals or groups on the machines and materials used and on the technique required.

Social Training.

Social life within the hostel forms an integral part of training. Many paradoxes are revealed, by study of the residents' behaviour, which show that conduct is often a manifestation of expediency. One well-groomed youth may be independent and have a well-developed dress sense. Another may have come from an over-protective home where he has been dressed and washed by his parents and never learnt to attend to his own toilet. An unkempt youth may have been neglected at home and had no social training; on the other hand, he may have had wise parents who have preferred to let him do everything for himself, even if badly and untidily, in order to acquire independence. None can be taken at his face value. They will not always be resident in their own homes, and the basis of social training must be the development of a good work-mate (as well as a good work-man) and one who could live independently and, perhaps, marry. This duty falls heavily on the house parents, but it concerns the whole staff.

All the cases of cerebral palsy we have admitted have been immature in the sense of poor achievement in formal education or social and cultural development. Deafness, dysarthria, and inability to write prejudice communication and the exchange of ideas. Immobility confines environment and largely prevents participation in team

games, excursions, formation of friendships with non-handicapped persons, and acquaintance with people of their own age. Limitation of circumstance to the family circle and the television screen prevents the development of powers of thought, expression, and judgment which derive from general conversation, handicrafts, and worldly experience. These deficiencies are remedied by evening classes in reading, writing and arithmetic and ballroom dancing, by study groups and lectures on current affairs, excursions to places of interest, and religious services. The response varies, but in general they are all receptive, and it is usual, for instance, for the reading age to advance from 5 to 15 years at the rate of a year a month.

Both work and social training have a therapeutic application complementary to the services of the physiotherapist and speech therapist. The physiotherapist's function is to correct contractural deformities and bad habits of posture and movement, to improve physique by exercises, and to apply specific remedial treatment after orthopaedic surgery. She also gives breathing and relaxation exercises to assist the speech therapist who deals with problems of phonation, articulation and dribbling.

Most cases of cerebral palsy have already had up to 25 years' 'treatment' by the time they enter, and they and their parents still look for a cure. However strong the temptation is to prescribe treatment on its own intrinsic merits, it has always to be remembered that time enough has already been devoted to this purpose, and at this age the importance and urgency of vocational training is paramount. Any physical or surgical procedure must be purpose designed in relation to work training. The object of work training must be towards productive work rather than work involving service and, as in universities as distinct from technical colleges, the attitude to

work must be that it is a way of life as well as a means to an end.

Some Principles of Work Therapy

A man with cerebral palsy cannot be trained to work unless the actual functional movements required are within his physical competence. It is no use expecting a hemiplegic with one effective hand to tie the knots of string bags or an athetoid with violent uncontrolled movements to make paper lamp-shades. Fingering, handling, and the use of hand-tools are far more difficult and more skilled actions than tending power-driven and manually operated machines. It is therefore necessary to reverse the usual scheme of apprenticeship training, wherein tuition in the theory of the subject, with instruction in bench work and the use of gauges, marking instruments, and hand-tools usually precedes work in the actual craft or trade. The man with cerebral palsy must start on unskilled, or semi-skilled, work, and, as his co-ordination and functional capacity are improved by constant repetition of controlled actions, he will progress to work involving less coarse movement. Standing or sitting posture must be corrected or adjusted to place the limbs in the best position of mechanical advantage in relation to the machine. Proper foot or leg supports are required by those who cannot plant their feet firmly on the ground. There is a great waste of effort when every push and pull of the arms twists the body on the seat in the absence of leg fixation.

In industry, limb movements may be straight or curved in any one of three planes which are mutually at right-angles. Pillar drilling and mortising machines both require an upward reach and downward pull of one hand and arm in the antero-posterior plane. Most people use their right hand to pull the lever and their left to position and steady the work. Problems of laterality and crossed-laterality exist for

some cases, and handedness needs study to secure the best productivity. There is, however, a general difference between the operation of these two particular machines which illustrates a factor influencing selection of training. Metal to be drilled is commonly in small pieces which can be picked from one tray, drilled, and handed to another tray within reach. Timber pieces, on the other hand, are usually bulky and have to be fetched to the machine and then carried away and stacked. Training in woodworking is best suited to those who are ambulant.

Press tools may be operated by hand or foot, but the commonest type involves pulling a vertical handle in a horizontal plane along the arc of a circle with a radius of up to two feet. Considerable effort is required to overcome the inertia of the machine and impart momentum to the system. Athetoids, having strength but lacking co-ordination, operate this machine well. The initial strenuous effort abolishes uncontrolled movement and, by its momentum, the moving handle carries the hand and arm through a controlled range of movement. Uncontrolled movements tend to decrease both with repetition and with the development of muscle fatigue. The reward of every operation is the conversion of purposeless movement to effective productive use.

Woodworking machines, such as circular saws, planers, thicknessers and sanders, require the material to be offered to, and fed through, the machine with a constant steady pressure. If the timber is of any length the operative will have to carry it, and walk with it, as it feeds through. The imposition of a constant effort, necessarily requiring no change in rate or force, conduces to abolition of unwanted movement in athetosis and, paradoxically, to controlled activity in purely spastic states.

Effort in the third plane, the coronal plane, is secured in working injection

moulders by means of a horizontally placed lever or a large vertical capstan wheel. Whereas presses require initial effort, the wheels and levers of these machines require increasing effort ending with a sustained force. This increases muscle stamina and, again, steadies those with unwanted movements.

These fundamental limb movements are still used in modified form and on a smaller scale in lathes, milling machines, sanders and grinders, and in the guillotines, shears and folders used in working sheet metal. As work therapy stimulates neuromuscular intergration and facilitation, the subject can progress from one class of machine to the next and on to occupations requiring handling and fingering. This class of work is to be found in the use of hand-tools for finishing, assembling light components, and packaging.

Printing.

Printing demands a high degree of manual dexterity, an ability to spell, an understanding of words, and keen visual perception in three dimensions. While setting up type in its frame ranks as assembling, and hand printing is essentially repetitive work, the whole process develops craftsmanship and affords a means of stimulating special traits of character such as artistry, concentration, accuracy and neatness. Valuable as this facility is for providing training in certain respects, it has to be realised that entry to the printing trade is exclusively by apprenticeship and this is a right guarded by the Trades Union. The situation has to be respected even if it means that printing is a trade consequently closed to all the cases of cerebral palsy we have seen. It follows that vocational training in printing, as distinct from its use therapeutically, is only valid if there is a possibility of eventual employment by an industrial firm with its own printing section.

Concentration.

For some obscure reason hemiplegics, as a type, commonly lack concentration. For other types of cerebral palsy time hangs heavily after they have left school, and they seem to relish tedious time-consuming tasks. Traits of character, such as lack of concentration and perseverance, and inability to sustain effort, are as much due to low intelligence as to physical weakness. Sheer laziness merits disciplinary action, but it needs a more subtle approach to overcome lack of ability to concentrate. The best results are achieved if the instructor can anticipate or soon observe wandering interest and attention, and can quickly introduce a diversion which is purposeful in itself. He can, for instance, ask the trainee to carry a message or fetch another tool, or to count production so far and calculate what would be a fair output for the day. Respite from work then ceases to be time wasting and gradually a habit of sustained effort is acquired.

Conclusions

It has been shown that, out of 87 cases admitted, 41 have been trained and placed in employment, 24 are still in training, and 22 have left the centre for various reasons. The apparent failure rate is 25 per cent. However, only 8 cases were discharged as untrainable for reasons directly attributable to cerebral palsy. The absolute failure-rate has therefore been 9 per cent. During the first two years we did not fully realise the impact the social behaviour of trainees and parents would have on vocational training, but this experience leads us to believe we can now meet it more adequately by change of approach to, and management of, individuals. The inference is that more than 75 per cent of the cases we select will be successfully trained and placed in open employment.

No two identical cases have been seen

and so far it is impossible to set down any criteria of trainability. The problem presented by each applicant has been a personal challenge to ourselves—what could we do with the individual with the facilities available? The first 236 cases referred to us were all officially regarded as unemployable and generally considered to be untrainable because they suffered from cerebral palsy; 67 cases were rejected at interview and 169 accepted for admission. Application of the apparent 25 per cent failure rate to the 169 acceptances indicates that 42 will leave the centre without being placed in employment. These figures indicate that 127 out of 236 cases—54 per cent—are trainable and will be placed in open employment. This figure is enhanced to 65 per cent if a 10 per cent failure rate is assumed.

This argument has been pursued to demonstrate that the work potentiality of the cerebral palsied, as a class, has been misjudged and under-rated. Achievements can become greater still, but not easily. The short answer would be to secure more promising material on which to work. This raises certain matters concerning education and medical treatment for discussion.

Comment has already been made on immaturity and backwardness in educational standards. Except for children with normal intelligence and only slight physical disability, it would seem that a school syllabus could be planned more realistically in relation to the likely life the child will lead after leaving school. More concentration is needed on practical subjects and the practical application of those subjects. In our experience with the 16-25-year age-group, the reading age often advances from 5 to 15 years at the rate of a year a month. This suggests that chronological age may well be disregarded in some cases, and the upper limits of primary and secondary modern education should be permitted officially well beyond

11 and 15 years respectively. Maturity and independence depend not only on intellectual attainments but also on an adequate and sufficiently varied community life. This does seem to have been too confined in those of this series of cases who have been in residential schools, and it certainly appears that devoted care and supervision of children can shelter them too much so that they leave school still dependent and immature.

A few cases in this series had no therapy of any description but most had been having treatment of one form or another all their lives. Those who had had none were at no disadvantage compared with those who had. In no single case has a parent attributed any specific benefit to physiotherapy, although those currently having treatment all wished for treatment to be continued at Sherrards.

In 98 cases surgical treatment had been given before interview. There were 8 cases of arthrodesis of the wrist or ankle, 2 of femoral osteotomy, and the remainder had all had numerous tenotomies and neurectomies. It is quite clear that the

hemiplegic is an easier surgical proposition than the paraplegic. It is also clear that those who have had a planned series of operations have derived more benefit than those who have had isolated tenotomies followed by second and third interventions carried out piecemeal. The impression created in the mind of a physician is that operation on an isolated joint is never justified if significant postural defects exist in other joints of that limb. Furthermore, the operations should proceed from the proximal to the distal joints. Some of the parents of applicants rejected because of inability to walk said that they had lost their ability to walk after surgical treatment. It does appear that the cerebral palsied have been operated on too often and at too early an age and that further experience will suggest that surgical intervention should be deferred, if possible, until body growth has ceased.

Acknowledgements: I wish to thank the National Spastics Society for permission to write this article and the Principal of Sherrards, Mr. E. L. Knight, for his constant friendship and co-operation.

SUMMARY

Three years' experience in training young adult cases of cerebral palsy is reported.

The method of assessment and selection of candidates is described and the main presenting features of 211 cases analysed.

A description is given of the facilities which exist and are projected at Sherrards Training Centre.

Reference is made to the social, domestic and technical problems presented by the trainees and an explanation is offered where the attempt at training has failed.

Comments are made on the education and medical and surgical treatment previously given to the cases examined.

RÉSUMÉ

Formation professionnelle pour infirmes moteurs cérébraux

Compte rendu de trois années d'expérience en matière de formation de jeunes adultes infirmes moteurs cérébraux.

Description de la méthode d'évaluation et de sélection des candidats, et analyse des principales caractéristiques de 211 cas.

Description des facilités existant et à l'étude au Centre de Formation de Sherrards.

Référence aux problèmes sociaux, domestiques et techniques présentés par les élèves, et explication des cas d'échecs.

Discussions sur l'éducation et le traitement médical et chirurgical reçus antérieurement par les sujets examinés.

ZUSAMMENFASSUNG

Berufs-bildung für Patienten mit Zerebrallähmung

Es wird über dreijährige Erfahrung im Gebiet der Ausbildung junger Erwachsener mit Zerebrallähmung berichtet.

Die Schätzungs- und Auswahlmethoden der Kandidaten werden beschrieben und die hauptsächlichsten Merkmale von 211 Fällen analysiert.

Man beschreibt die vorhandenen und geplanten Erleichterungen in der Ausbildungsanstalt von Sherrards.

Man verweist auf die sozialen, häuslichen und technischen Probleme, die die Schüler stellen und bringt eine Erklärung der Misserfolge.

Man gibt Erläuterungen über die Erziehung und die medizinische und chirurgische Behandlung, die die untersuchten Patienten vorker erhalten hatten.

NOTICE

Congresses in Ankara and Lisbon

THE Second Middle-East-Mediterranean Paediatric Congress will be held in Ankara from September 6 to 9, 1961. Brochures are available from the Hon. Sec. British Paediatric Association (Dr. E. W. Hart, M.B.E., F.R.C.P., at the Institute for Child Health, The Hospital for Sick Children, Great Ormond Street, London, W.C.1).

The Tenth International Paediatric Congress will be held in Lisbon from September 9 to 15, 1962. Those proposing to attend should register between August 1, 1961 and January 31, 1962.

The Development of Visual Capacity in the Infant and Young Child

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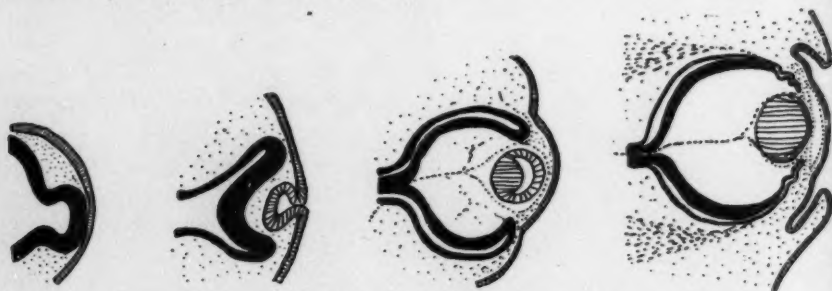
Development of the Eye

A BRIEF description of the development of the eye may help one to understand its condition at birth. At the third week of embryonic life (3-4 mm.) the eye starts to develop from two lateral outpouchings at the end of the neural tube in the region of the forebrain (Fig. 1A). The neck of the vesicle narrows to form the optic stalk (the future optic nerve). A secondary invagination of the vesicle starts on its under-surface in the foetal fissure and

proceeds to involve the whole surface, thus forming the optic cup (Fig. 1B). The overlying ectoderm thickens to form the lens plate. This grows inwards and gets cut off from the surface, thus forming the embryonic lens (14 mm., 6 weeks) (Fig. 1C).

The *hyaloid artery*, a branch of the ophthalmic artery, grows into the foetal fissure and forward to the back surface of the lens, where it forms a vascular network on its posterior and lateral surfaces. The ophthalmic artery continues forward to

Fig. 1. A, B, C, D. Normal development of the Human Eye (after Mann). Neural ectoderm in black. Surface ectoderm shaded. Mesoderm dotted. This figure and Fig. 2 are taken from Duke-Elder's Text book of Ophthalmology, by kind permission of the author and the publishers, Henry Kimpton.



A—Primary optic vesicle, seen as an outgrowth of forebrain in contact with surface ectoderm.

B—Commencing invagination of primary optic vesicle and appearance of lens pit in surface ectoderm.

C—Deepening of optic cup, separation of lens vesicle from surface and appearance of hyaloid blood system inside the eye.

D—Commencing forward growth of margin of optic cup to form ciliary region and ectodermal part of iris. Formation of lens fibres from cells of posterior wall of lens vesicle. Appearance of anterior chamber and lid-folds. Mesoderm condensing around.

the rim of the optic cup, where it becomes the *annular* vessel, and, anastomosing with the hyaloid, forms the anterior vascular tunic of the lens (pupillary membrane). By the 9th week the hyaloid system is at its maximum, filling the cavity of the optic vesicle (the primitive vitreous). It then starts to atrophy, but remnants of it are nearly always visible in small premature infants (under 32 weeks' gestation). Atrophy occurs from before backwards, the posterior half of the hyaloid artery being the last remnant.

The anterior margin of the optic cup, which has grown forwards towards the lens, forms the *ciliary* region and the ectodermal layers of the *iris* (Fig. 1D). The inner layer of the optic cup forms the neural layer of the *retina*, and the outer becomes a single layer of pigment cells. Retinal development is well advanced at an early stage, there being two distinct layers as early as the 6th week (14 mm.). The *retinal vessels* develop from the hyaloid vessels at 15 weeks but do not reach the ora serrata till the 9th month. The rest of the eyeball (cornea, sclera, anterior layers of iris, ciliary muscle) are all developed from paraxial mesoderm.

Anatomy in the Premature Infant

Globe: In the full-term infant the eyes, together with the brain, are more fully developed than the rest of the body. This difference is greater in the premature infant. At 7 months the eyes appear to be too large for the orbits, and there is often an appearance of exophthalmos, only the equator of the globes being covered by the orbital margins. In addition, the orbital entrance is circular and the orbital margins are more closely applied to the globes than in the full-term infant.

Vascular System: In the small premature infant (less than 32 weeks gestation) the vascular network in the pupil (anterior tunica vasculosa lentis) and the hyaloid

artery stretching from disc to lens are usually visible with the ophthalmoscope. The pupillary network is best seen with a + 12 D lens in the ophthalmoscope, and the hyaloid artery can be followed in its course with a lens varying from - 10 D to + 12 D.

At this stage of prematurity the view is hazy because of the poor translucency of the media, and the eye is usually myopic. (The myopia is temporary and disappears in most cases by 3 months of age.)

The persistence of these foetal remnants is much commoner in the small premature infants. Schmöger (1955) found the pupillary membrane constantly present in the first week of life in babies weighing under 2.2 lb. at birth, in 86.5 per cent of those weighing 2.2-3.3 lb., in 74.5 per cent of those weighing 3.3-4.4 lb., and in 40 per cent of those weighing 4.4-5.5 lb. at birth. Similarly, Roper-Hall (1960) found the hyaloid artery in 92 per cent of those weighing 2-3 lb. at birth, in 58 per cent of those weighing 3-4 lb., in 36 per cent of those weighing 4-5 lb., and in 13 per cent of those weighing 5-6 lb. at birth. The pupillary membrane usually disappears in the first week, but in the small prematures it may persist for several weeks. They hyaloid artery persists longer, but has usually been absorbed before the infant reaches his full-term date. The posterior half is the last to go, tending to coil in front of the disc before it disappears.

The Anterior Segment of the eye has a small cornea, a shallow anterior chamber, a small pupil, and a structureless iris of a homogeneous grey-blue colour. The lens is spherical.

The Fundus Picture

(a) The *disc* has a greyish pallor which persists for several months. At this stage it is impossible to say if optic atrophy is present or not.

(b) The *fundus* looks pale, and there are white patches round the disc-macula area

where sclera is visible because the choroidal capillary network and choroidal pigment layer is incomplete. There is a wide grey-white area in the periphery of the fundus where neither choroidal nor retinal circulations have yet developed. The more premature the infant, the wider is this area.

(c) The *macula* is very immature. It is thicker than the rest of the retina because it contains a large number of ganglion cells, several layers deep, and the fovea is not yet hollowed out (see Fig. 2A). The cones are short and poorly developed. These structural conditions explain the lack of fixation in the small premature infant at birth.

Refraction: In the small premature infant, the hazy ophthalmoscopic view is

often associated with a temporary myopia varying from 0 to 12 dioptres. This usually disappears by 3 months, but occasionally persists till 6 months. It has not yet been determined whether this is due to variations in curvature of cornea and lens or to a refractive difference in the media.

This must be distinguished from the myopia that is a manifestation of mild Retrolental Fibroplasia, which is a non-progressive but permanent myopia.

Anatomy in the Full-Term Infant

Globe: The orbits have now grown larger relative to the eyes, so that the exophthalmic appearance of the premature infant has disappeared. The orbital margins are also less closely applied to the globes. They are stronger and are becoming oval.

Vascular system: The primitive vascular system, consisting of the hyaloid artery and pupillary membrane, has disappeared and the media are now perfectly clear.

The Anterior Segment is more like the adult eye than it is in the premature infant. The cornea is nearly of normal size but flatter than in the adult. It is fully grown by 2 years of age. The anterior chamber is less shallow than in the premature infant, but still shallower than in the adult. The pupil is small and the iris still a structureless grey-blue. It does not develop its delicate meshwork and adult colour till the baby is a few months old.

The Fundus Picture is more like the adult eye, although some differences remain.

(a) The *disc* has a greyish-white pallor at birth. As medullation of the optic nerve fibres develops in the first few weeks after birth, the disc becomes white rather than grey. Gradually the capillaries ramify in its substance, and by 2-4 months it attains its normal pink colour.

(b) The *choroidal capillary network* and pigment have now fully developed and the

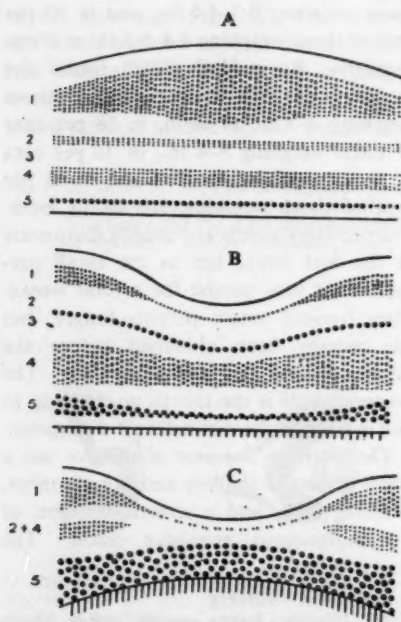


Fig. 2. Development of the macula (after Mann), showing the cell layers (A) at 6 months of foetal life; (B) at birth; and (C) in the adult.

- | | |
|-------------------------|-----------------------|
| 1. Ganglion cell layer. | 3. Layer of Chievitz. |
| 2. Amacrine cells. | 4. Bipolar cells. |
| | 5. Nuclei of cones. |

fundus has the uniform red background of the adult. There is still a small peripheral white zone, but this is much narrower than in the premature infant.

(c) The *macula* (Fig. 2B) is better developed than in the premature infant but still has not attained adult form (Fig. 2C). The ganglion cell layer has become thinner by the migration of the ganglion cells away from the fovea. The latter therefore becomes hollowed out and light is allowed more direct access to the cones. At birth these are still short and ill-developed, but in the first 4 months they gradually develop their adult form, becoming larger and more numerous, and their nuclei becoming 3 layers thick. Thus the baby is 4 months old before the macula has fully developed.

Refraction: The full-term baby is usually hypermetropic at birth (Duke-Elder 1949), the average being about +2 Dioptres. This hypermetropia tends to increase slightly between birth and 7 years of age. As the eyeball grows longer there will be a tendency to myopia, but this is offset by a decrease in the refracting power of the lens. At birth it is nearly spherical but with growth it becomes more lenticular (biconvex). (An almost spherical lens may account for the myopia of the premature infant.)

Physiology

Fixation of Light involves a reflex path, in which the afferent pathways are: retina, optic nerve and tracts, lateral geniculate body, optic radiation, and occipital cortex; while the efferent are: occipital cortex, oculomotor nuclei, oculomotor nerves, and extra-ocular muscles. Thus co-ordination of the 6 extra-ocular muscles in each eye is required immediately after birth to carry out reflex fixation of light. This the full-term infant attempts in the first few days of life and the premature infant a few weeks later. Owing to the late develop-

ment of the macula, the baby's visual acuity in the first few weeks is poor, so these attempts at fixation can be made only if a bright light is held close to the eyes.

The *Local Sign* of the retina is an innate function whereby the infant can project impulses falling on the retina to a particular part of his visual field and thus later can recognise the direction from which they come. So light shone from the temporal side on the right eye will stimulate the nasal retina, and there will be an involuntary movement of the right eye outwards towards the light. The impulses follow the reflex path described above. As the thinking processes gradually start to develop, the infant comes to learn that objects which stimulate the left side of the retinae lie on his right and vice versa.

Voluntary fixation of an object involves a more complex neural pathway which includes a relay of impulses from the occipital cortex to the frontal lobe (second frontal gyrus) and thence to the oculomotor nuclei. This does not usually develop until 1 or 2 months.

Further, visual sensations have to be related to many other sensations received from other organs—taste, touch, hearing, balance, etc. Since these are all correlated in the brain with motor activity, the infant gradually builds up a composite picture of the new world into which he has been born.

Binocular Vision. For the first 6 weeks the infant has been using monocular vision, making little or not attempt to use the eyes together. Binocular vision cannot be present to any significant extent until the maculae have fully developed. Nor can it occur until co-ordination has developed between the extra-ocular muscles of the two eyes. It will be remembered that each of the 6 extra-ocular muscles works in conjunction with one muscle in the opposite eye which contracts with it—the

contralateral synergist—and one muscle in the same eye, the ipsilateral antagonist, which relaxes as the first muscle contracts. These muscles carry out the greater part of the movement, but all 12 extra-ocular muscles are concerned to some degree in any ocular movement. The external and internal recti are responsible for horizontal movements, the superior recti and inferior obliques for elevation, and the inferior recti and superior obliques for depression. The vertical recti have their maximal action when the eye is abducted, while the obliques have theirs when the eye is adducted. Thus on dextroversion the right lateral rectus contracts with the left medial rectus (through reciprocal innervation) and the antagonists (right medial rectus and left lateral rectus) relax (see Fig. 3). Similarly, on looking up to the right the right superior rectus contracts together with the left inferior oblique, while their antagonists, the right inferior rectus and the left superior oblique, relax.

Deviations from the straight are common in the early months, though they do not occur in all babies. When they are intermittent they can be safely discounted. Co-ordination of the eyes is learnt gradually from 6 weeks onwards. Lapses from the straight position become less frequent as binocular vision becomes stronger and by 9 months the eyes should be straight all the time.

Methods of Examination

In the very young infant, the simplest test is to shine the light of a torch on one eye at a time in a semi-darkened room. This test is done to see whether the infant attempts fixation and if so through what range of movement fixation can be held. Large objects like a 6-inch black ring or a large brightly coloured toy can also be dangled in front of his eyes and his attempts at fixation can be assessed. As he

grows older, he should be able to pay attention to smaller objects.

Another simple test is the Doll's Eye Phenomenon (Mac Keith 1961). An infant is wrapped in a blanket and held up and turned round. In the early days the eyes lag behind the direction of movement, but in later weeks the eyes will move in the direction of movement and will fix a bright window before the face is turned to it.

An apparatus for testing infants' vision designed by Schwarting (1954) consists of an illuminated box with a pointer which moves to and fro against the bright back-

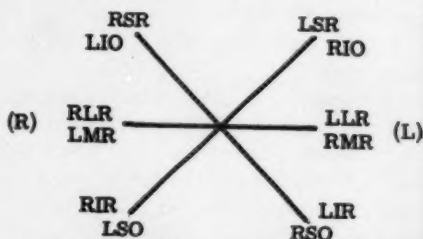


Fig. 3 The directions in which the extra-ocular muscles exert their maximal action.

ground. The infant's eyes are watched to see if they follow the pointer, the size of which can be reduced or increased as required.

Examination of the fundi and media is essential in any assessment of visual acuity. This should be carried out in a darkened room 20–30 minutes after the instillation of 1 per cent homatropine drops. The examination is much easier to do when the infant is sucking a feeding-bottle. Sucking relaxes the orbicularis oculi, and the lids can then easily be held open with the examiner's fingers. Sometimes the nurse can help by gently drawing down the lower lid while the examiner, from above the baby's head, holds the upper lid with one hand and the ophthalmoscope with the other.

Visual Capacity in the Premature Infant

At 28-32 weeks: The small premature infant has his eyes closed for the first week or two after birth. He makes no attempt to fix light. However, his eyes are sensitive to light, and if a bright light is shone on them he may make a sharp avoidance response. He will start opening his eyes and begin to look round well before he reaches his full-term date. Indeed, he may do so as early as 2 weeks after birth though he may be quite normally not do so till 5 or 6 weeks. At this stage of prematurity the eyes often look exophthalmic owing to the relative smallness of the orbits, referred to in the section on Anatomy in the Premature Infant.

At 32-36 weeks: Babies born at this stage will show more dislike for very bright light than the smaller ones and will be quicker in following the light of a torch. Thus a baby born at 34 weeks will probably be opening his eyes and taking notice by 2 or 3 weeks. When born at 36 weeks he will attempt momentary fixation of a large object dangled in front of his eyes. The attempt is fleeting and cannot be called true fixation, but a crude form of fixation does develop in the first 2 weeks.

Visual Capacity in the full-term Infant

At birth: The infant usually keeps his eyes closed for the first few days of life, but some start looking round a few hours after birth. Crude fixation is present, and he will follow a light or a large object, such as a black 4-inch ring, dangled less than 2 feet from his eyes against a light background. This fixation is usually monocular and is limited to 45° of arc or less (Keeney 1951). Some photophobia is still present and he will close his eyes immediately if a bright light is shone close to them.

At 2 weeks: Monocular fixation gradually improves, and he will now

follow the 4-inch black ring up to 3 feet from his eyes. A bright light shone in the peripheral field of one eye will attract his attention and his eye will move towards it. Vision at this stage is poor and is confined to large objects close to the eyes. It must be stressed that the world of the baby in the first few weeks of life is a world of near large objects only and he cannot see distant objects at all.

At 4 weeks: Vision is still monocular, with rapid alternation between the eyes. Monocular fixation has improved, and he can now follow the 4-inch black ring with head and eye through 90° of arc. He will now look at his mother's face when she speaks to him within 2 or 3 feet but not if she is further away.

At 6 weeks: The first sign of binocular vision now appears, and brief periods of binocular fixation of large near objects are attempted. This is, of course, an average time and many babies are earlier or later in doing this. Some, indeed, appear to use their eyes together from birth—apart, of course, from the spasms of convergence when they have wind. If the baby loses sight of a slowly moving object, he will now attempt to regain fixation. At this stage, too, he gives his first social smile on seeing his mother's face.

At 2 months he starts to pay attention to objects up to 6 feet away. He will look at the examiner's hand and will be especially attracted by a source of light like a window. At this stage the ocular movements on attempted fixation are still jerky.

At 3 months he can follow moving objects with head and eye movement through the whole visual field (180°), and the movements are becoming smoother. Visual acuity is improving, and he will take glances at smaller objects around an inch in diameter, though he prefers to

look at the hand that is holding it or the face near it. Voluntary convergence is just beginning, but it is very weak and fleeting. At this stage he begins to look at his own hands. Binocular vision is improving. This is consistent with improved visual acuity as the maculae near their full development (see above) and as the co-ordination of the ocular muscles improves with experience. Monocular fixation is now becoming less frequent and binocular fixation is becoming the rule.

At 4 months: Visual acuity has further improved and is now about 3/60 (i.e., equivalent to seeing the top letter of the test type at 3 metres). He is still more interested in near objects and will take some interest in small bright objects like a red golf tee. He will give prolonged regard to his rattle, which he can hold but cannot yet retrieve if he drops it. Binocular vision is still improving and lapses into

monocular fixation are becoming rare. Voluntary convergence is a lot stronger and can be maintained for a short time by toys held close to his eyes (Fig. 4).

At 5 months: Co-ordination of hand and eye is beginning to develop and he will start making efforts to grasp a rattle with some success. This improves between 5 and 6 months. Fig. 5 shows the baby grasping his toy with both hands quite successfully. The attempts are at first clumsy raking movements, and are successful only if the object is large and well within reach. Co-ordination of eyes and ears is also developing. Thus, if a noise is made somewhere in the room, he will look round towards it.

At 6 months: Visual acuity has improved to approximately 4/60 and he can now recognise faces up to 6 yards away. He watches the Television picture from 3 yards, even when no sound is on. He reaches out



Fig. 4. Voluntary convergence in a baby of 4 months.



Fig. 5. Baby of 5½ months reaching out to grasp a toy with both hands.

for objects near him and is more successful in picking them up, being able to retrieve a dropped rattle and to pick up objects such as a 1-inch red cube. At this stage he will look with interest at his image in a mirror and will try to touch it. He will also adjust his position to see people by craning his neck backwards or forwards. Binocular vision is stronger and he resents the diplopia caused by a vertical prism placed in front of one eye. Lapses into squint are now rare.

At 9 months: Further improvement in all visual achievements has occurred. Visual acuity is better and he can see tiny pieces of white paper 2 or 3 mm. in diameter lying near him. Co-ordination between hand and eye is better, too, and he can pick up the tiny piece of paper referred to with a neat pincers movement. He looks intently at expressions on faces and attempts to respond with a similar expression. His ocular movements are smooth in both horizontal and vertical directions of gaze.

At 1 year: Visual acuity is now at least 6/60 and may be as good as 6/24. Certainly his near visual acuity is very good.

Binocular vision is stronger and some weak fusion is probably present, although this is impossible to estimate at this age. Co-ordination of eye, hand and ear is improving all the time.

At 2-5 years. By 2 years his visual acuity is 6/12, by 3 years 6/9 and by 5 years 6/6. Stronger fusion and some stereoscopic vision is present by 3 years, and by 4 years good stereopsis should be present.

Gradually the child's world is widening out. In the early weeks he is aware of little except his mother's face within 1 or 2 feet. After 2 or 3 months he begins to be aware of other members of the family, but only when their faces come within 2 or 3 feet. At first his world is an indistinct near world and nothing more distant is visible. By 6 months his horizon is widening and he is aware of things happening across the room. Between 6 and 12 months it widens still further and he begins to realise that there is an outside world beyond his home. Gradually he begins the fascinating process of learning about it through his eyes—a process that will continue all his life.

SUMMARY

A brief description is given of the development of the eye in utero, with special reference to the embryonic structures which persist in premature babies.

The anatomical features which distinguish the eye of the premature and full-term infant from that of the adult are outlined.

The physiological mechanisms involved in the development of vision in the infant are discussed.

The methods of examination of babies' eyes are briefly described.

The development of visual capacity in the premature and full-term infant is followed through the early months of life, and an attempt is made to assess the visual acuity at each stage.

RÉSUMÉ

Developpement de la capacité visuelle chez le nourrisson

Nous trouvons dans cet article:

une brève description du développement de l'oeil in utero avec référence particulière aux structures embryonnaires persistant chez les prématurés;

un aperçu des signes anatomiques distinguant l'oeil de l'enfant prématuré et à terme de celui de l'adulte;

une discussion sur les mécanismes physiologiques mis en jeu dans le développement de la vision chez le nourrisson.

Les méthodes d'examens de l'oeil du petit nourrisson sont décrites.

Le développement de la capacité visuelle chez le prématuré et l'enfant à terme est suivi pendant les premiers mois de la vie, cependant que l'on cherche à évaluer l'acuité visuelle à chaque étape du développement.

ZUSAMMENFASSUNG

Entwicklung der Sehfähigkeit bei dem Säugling

Die Entwicklung des Auges in utero wird kurz beschrieben, mit spezieller Verweisung auf die embryonnären Strukturen die bei Frühgeborenen fortbestehen.

Die anatomischen Unterscheidungszeichen zwischen dem Auge des frühgeborenen und ausgetragenen Kindes und dem des Erwachsenen werden kurz besprochen.

Die physiologischen Mechanismen die die Entwicklung der Sehfähigkeit des Säuglings in sich schliesst, werden erörtert.

Die Untersuchungsmethoden des Auges des Säuglings werden kurz beschrieben.

Die Entwicklung der Sehfähigkeit bei dem Frühgeborenen und dem normal ausgetragenen Kinde wird während der ersten Lebensmonate gefolgt und man versucht, die Sehschärfe in jedem Stadium abzuschätzen.

REFERENCES

- Duke-Elder, S. (1949) Textbook of Ophthalmology, Vol. 4, p. 4263. London: Henry Kimpton.
Keeney, A. H. (1951) Chronology of Ophthalmic Development. Springfield, Ill.: Thomas.
Mac Keith, R. (1961) 'The baby who may be blind,' *Trans. ophthal. Soc. U.K.* 80, 13
Roper-Hall, M. J. (1960) 'The eye of the premature baby.' *Brit. med. J.*, ii, 231.
Schmöger, R. (1955) 'Die Pupillarmembran als Zeichen der Unreife.' *Kinderärztl. Prax.*, 23, 433.
Schwartz, B. H. (1954) 'Testing infants' vision. An apparatus for estimating the visual acuity of infants and young children.' *Amer. J. Ophthal.*, 38, 714.

The Development of Myopia in Early and Later Childhood

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MYOPIA virtually always develops in childhood or adolescence, though its complications, which may lead to blindness, usually appear in early middle age or later. The criteria which, at an early age, distinguish a case which will deteriorate progressively and remorselessly to retinal degeneration and blindness from one which will merely become intermittently worse but never reach a disabling level are not well established. It is, however, the rule for the unaided distance vision of the majority of myopic children to deteriorate to some extent once the process begins until growth stops. Since myopia is the commonest cause of blindness in the prime of life and is responsible for considerable visual anxiety in many other people both adult and younger, the condition has prompted a considerable effort of research from time to time in various parts of the world, but little progress has been made in understanding it. It is agreed that myopia is a disease of growth and development, and it is logical to make serious attempts at its prevention and control during childhood or even antenatally.

Nature of the Condition

In its simplest terms the myopic eye is too long anteroposteriorly to enable rays of light from distant objects to be focussed on the retina. Theoretically this can be due to an increase in the actual anteroposterior diameter or to an increase in power—i.e., curvature—of the cornea, or to an increase

in power of the lens, or to combinations of these factors (Fig. 1). In spite of many attempts to clear up this optical problem it is still uncertain exactly what the typical optical lesion is in these terms, except that it is agreed that in myopes with errors of more than 4 dioptres there is always an increase in the actual anteroposterior diameter of the eye. (Usually this corresponds with A in Fig. 1). In the lower degrees of myopia there appears to be no characteristic optical measurement which is constantly present and the condition seems to be one of lack of co-ordination to which reference will be made later. The other characteristic of myopic eyes with more than —4.0 dioptres is that the sclera, choroid, retina and cornea are likely to be thinner than in a normal eye, which constitutes a pathological physical state and is the precursor of the degenerative changes leading to blindness.

Chronologically there appear to be two types of myopia—congenital and acquired—the former being rare, usually of a high order and non-progressive, while the latter is common and almost invariably progressive to some extent. It will be seen straight away, therefore, that these two types of myopia differ in their behaviour. It is not known whether the high non-progressive myopia of infants is more serious ultimately than the high myopia reached progressively in the acquired type.

There is undoubtedly an abnormally high proportion of short-sighted infants among those born prematurely (Drillien

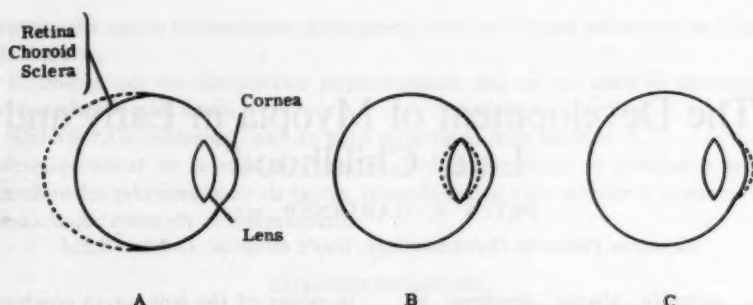


Fig. 1. Possible mechanisms of myopia. A: Increase in anteroposterior diameter of the eye. B: Increase in power of the lens. C: Increase in curvature of the cornea.

1958, Gregory 1957), though many figures reported from time to time are based on ophthalmoscopy and not on formal refraction. Such figures tend to give a fallaciously high estimate of the number of myopes and probably of the variability of the myopia in any given case. If the problem is approached in reverse there is a considerably higher incidence of maternal disease during pregnancy, especially of toxæmia, among the mothers of congenital myopes than among the mothers of normally sighted children. In these children prematurity is not a feature (Gardiner and James 1960).

Further unpublished studies of mine, which are being repeated, indicate that both prematurity and post-maturity are associated with either immediate or subsequent visual defects in about equal numbers.

Mongol children appear to have a susceptibility towards myopia in early childhood, which is probably congenital. About 30 per cent of them are myopic.

Therapy

In terms of treatment, visual education and general progress, highly myopic infants should start to wear glasses during their third year. The symptoms of myopia in these children are inconspicuous, and, since refraction under a mydriatic is simple and harmless in children of two,

this test should always be performed on children at this age where their birth-weight was under 4 lb., whether or not the child appears to have a visual defect. This not only settles the matter once and for all but also prevents some children growing up amblyopic in one eye whose symptoms, if any, are completely masked by the usefulness of the good eye. It is open to doubt whether the provision of glasses prevents an increase in the myopia but it certainly produces the optimum conditions for the full development of visual acuity.

Pathogenesis

Considering how common acquired myopia is, surprisingly little is known about its pathogenesis although theories, usually unsubstantiated, are many. Even the incidence of the condition is unknown, nor is it known whether or not it varies from time to time.

Heredity appears to pay a negligible part in the pathogenesis of congenital myopia. The association of myopia with retrolental fibroplasia is also probably fortuitous in that prematurity is common to both (Wagner 1957), but in view of some findings in acquired myopia further research is necessary to establish this. With our present knowledge, prevention of congenital myopia would be much nearer if all pregnant women were healthy and

parturition occurred when the baby was of normal weight.

French figures (Benoit 1958) seem to show that acquired myopia is more common in urban than in rural children, and in England the incidence in a London suburb was roughly $2\frac{1}{2}$ times that in rural Cambridge-

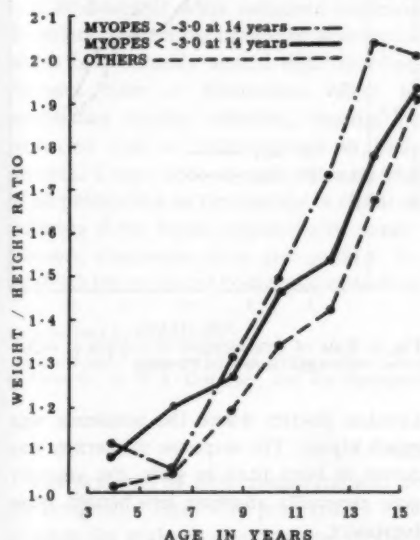


Fig. 2. Ratio of weight in lb. to height in inches in severely myopic, less myopic and normally sighted boys, aged 4-15 years.

shire (to be published). The disease commonly starts about the age of 7 years, and new myopes appear at each age from then on until about 15 in girls and 18 in boys. Whether ultimately there are more males or more females affected is unknown and the evidence is conflicting and insufficient.

The normal eye shows a finely governed co-ordination of corneal curvature, lens size and anteroposterior diameter. It is on this co-ordination during growth that normal vision depends. It is uncertain where this co-ordination breaks down in myopic eyes but this breakdown certainly does occur and in most cases can never be resolved. It is interesting that this lack of co-ordination in the growth of the

components of the eye occurs in children whose somatic growth is also different from that of other children and the degree of breakdown—i.e., the amount of myopia—appears to correlate to some extent with the amount of difference in physical growth in these children.

Figs. 2 and 3 illustrate these points. Children who become more myopic by the age of 14 have a higher weight/height ratio than those with rather less myopia, and both have a higher ratio than normally sighted children (Fig. 2). Secondly, weight increases are characteristically more irregular in myopes than in normally sighted children, though the pattern is the same (Fig. 3). Myopic girls tend to reach menarche earlier than the normally sighted.

It is interesting to note here that the appearance of myopia in young adults or its sudden deterioration at this age is

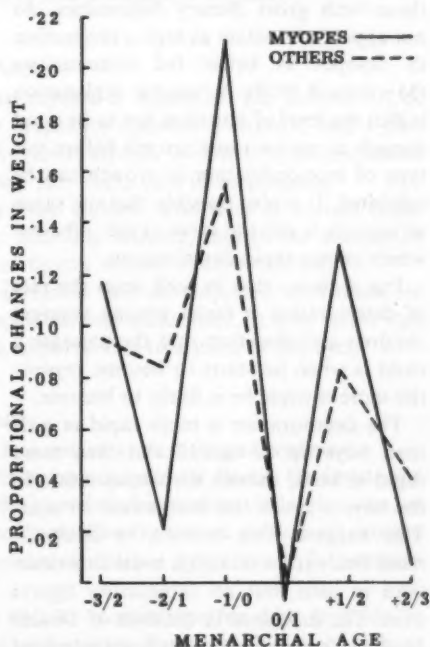


Fig. 3. Proportional rate of weight increase per menarchal year in myopic and normally sighted girls.

characteristically accompanied by equally sudden and considerable changes in body weight.

Whether these differentials in growth of the eye and somatic growth are linked or independent of each other, or depend on a common factor or factors is unknown. But there is one peculiar characteristic of myopic children, that they tend to eat less well balanced diets than normal children, and the assimilation of larger quantities of protein, particularly animal protein, does have a beneficial effect in slowing the rate of deterioration in their vision (Gardiner 1958). This indicates that myopia in the growing child is not entirely a genetic problem but can be dealt with in terms of the child's environment, and the observations should encourage further research.

A puzzling feature of this line of approach is that starving populations, or those with gross dietary deficiencies, do not appear to contain as high a proportion of myopes as better fed communities (Macdonald 1959). A possible explanation is that the level of nutrition has to be good enough to permit rapid growth before this type of inco-ordination of growth can be exhibited. It is also possible that the cause of myopia is not the same as the influence which causes rapid deterioration.

Fig. 4 shows that in both sexes the rate of deterioration is faster among younger children and therefore that the younger a child is when he starts to become myopic the more myopic he is likely to become.

The deterioration is more rapid in girls than boys up to age 13 and then more rapid in boys, though the deterioration in the boys is much less than at earlier ages. This suggests that myopia in adults is more likely to be of a high order in women than in men, but no satisfactory figures exist. The incidence in children of 14 and 15 shows a statistically significant excess of females in rural Cambridgeshire, but no difference between males and females in a

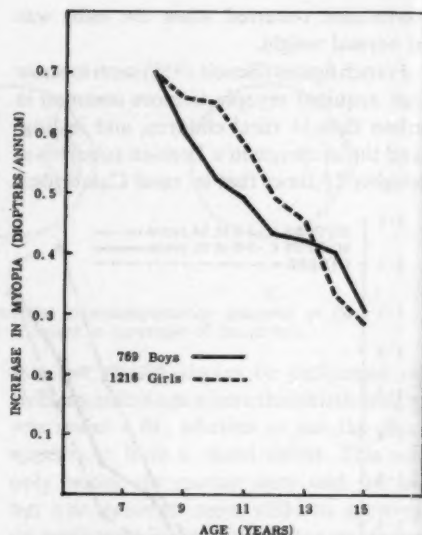


Fig. 4. Rate of deterioration of myopia at different ages from 8 to 15 years.

London district where the incidence was much higher. The response to therapy was better in boys than in girls, the younger girls especially showing less benefit from treatment.

These differences suggest a possible line of investigation and support the idea that myopia is due to a complex of factors.

In children with general physical handicaps, a preliminary survey shows that the incidence of myopia probably varies widely, possibly correlating with the type of somatic disease from which the child is suffering. In a series of children with cerebral palsy which I have recently examined myopia was considerably rarer than among the children with other gross physical handicaps—when prematurely born children were excluded (to be published)—and probably rarer than in the healthy child population. One could argue that as these children neither grow excessively quickly nor mature especially early, it is not surprising that few of them acquire myopia.

In other chronic disabling diseases, such as rheumatic fever and poliomyelitis, myopia appears to be much more common—probably commoner than in the population generally—but these figures require verification before they can be regarded as more than a basis for further research.

It is doubtful what influence systemic disorders have on the incidence of myopia, but it is commonly agreed that disabling illnesses have a deleterious effect in established myopic children, causing an increased rate of deterioration in their myopia. I have seen myopia increase out of all proportion to the average in children suffering from burns, appendicitis, tonsillectomy, rheumatic fever and pyelitis. To establish these clinical findings in individual

cases as having a genuine association in all cases would require a special investigation which it is hoped to undertake soon.

Conclusion

This brief description of myopia shows that it is a puzzling disease with its origins, its anatomical mechanisms and its treatment still largely a matter of guesswork; but none the less that further research is clearly necessary and likely ultimately to lead to a better understanding of the various factors implicated. Once these are elucidated and prevention in childhood rather than treatment becomes a possibility, there will be a reduction in the number of people with visual distress in later life.

ACKNOWLEDGEMENT:

Figs. 2 and 3 appeared in an article in *The Lancet* of March 6th, 1954, entitled 'The Relation of Myopia to Growth', by P. A. Gardiner, and are reproduced here by kind permission of *The Lancet*.

SUMMARY

The problem of both congenital and acquired myopia is described. The association of the congenital form with prematurity and with pregnancy disorder is stressed, and a plea is made for early recognition and treatment. Acquired myopia is shown to be associated with irregular somatic growth, and nutrition may play some part in its progress.

The incidence of myopia is low in cerebral palsied children who were of normal birth-weight but high in children with other chronic disabling diseases, though the absence of any normal standard of comparison makes these figures subject to confirmation.

This is a big problem, in which aetiology, incidence and development, and therefore prevention and treatment are inadequately mapped out and understood. It now seems that in many cases treatment of the child may be as important as treatment of the eyes.

RÉSUMÉ

Développement de la myopie au cours de la première et de la seconde enfance

Le problème de la myopie congénitale et de la myopie acquise est étudié. On insiste sur l'association de la forme congénitale avec la prématurité et des troubles de la grossesse, et l'auteur fait un plaidoyer en faveur d'un diagnostic et d'un traitement précoce. Il montre que la myopie acquise est associée à une croissance somatique irrégulière et la nutrition peut jouer un rôle dans son évolution.

L'incidence de la myopie est faible chez les enfants encéphalopathes dont le poids de naissance était normal, mais élevé chez ceux qui sont atteints d'autres maladies chroniques ou infirmités, bien qu'en l'absence de tout critère de comparaison standardisée chez des normaux, ces données demandent à être vérifiées.

C'est là un vaste problème dont l'étiologie, l'incidence et le développement et ce partant la prévention et le traitement sont encore insuffisamment esquissés et compris. Il semble maintenant que dans de nombreux cas, un traitement général de l'enfant soit au moins aussi important que le traitement de sa vue.

ZUSAMMENFASSUNG

Entwicklung der Kurzsichtigkeit im Frühen und späteren Kindesalter

Das Problem der angeborenen und erworbenen Kurzsichtigkeit wird dargestellt. Die Verbindung der angeborenen Form mit Frühgeburt und Störungen der Schwangerschaft wird betont und es wird eine Fürsprache für frühzeitige Diagnose und Behandlung gemacht. Es wird gezeigt, dass erworbene Myopie mit unregelmässigem körperlichen Wachstum verbunden ist und dass die Ernährung eine gewisse Rolle in ihren Fortschritten spielen kann.

Das Vorkommen von Myopie ist selten bei Kindern mit Zerebrallähmung, deren Geburtsgewicht normal war, aber häufig bei Kindern mit anderen chronischen verkrüppelnden Krankheiten, obgleich der Mangel an jeglichem Vergleichsmaßstab bei Normalen eine Bestätigung dieser Dase notwendig macht.

Es handelt sich hier um ein bedeutendes Problem, dessen Ätiologie Vorkommen und Entwicklung und folglich Vorbeugung und Behandlung unzulänglich dargelegt und verstanden werden. Es scheint jetzt, dass in vielen Fällen eine Behandlung des Kindes ebenso wichtig sein kann, wie eine Behandlung der Augen.

REFERENCES

- Benoit, A. (1958) 'Biotypologie del' homme myope.' *Arch. Ophtal., Paris*, 18, 734.
Drillien, C. M. (1958) 'Growth and development in a group of children of very low birth weight.' *Arch. Dis. Child.*, 33, 10.
Gardiner, P. A. (1958) 'Dietary treatment of myopia in children.' *Lancet*, i, 1152.
— James, G. (1960) 'Association between maternal disease during pregnancy and myopia in the child.' *Brit. J. Ophthalmol.*, 44, 172.
Gregory, I. D. R. (1957) 'Retinopathy of prematurity (retrolental fibroplasia) in children in whom the disease has not progressed to complete blindness.' *Ibid.*, 41, 321.
Macdonald, I. (1959) 'Visual acuity of South African undergraduates.' *S. Afr. J. med. Sci.*, 24, 1.
Wagner, G. (1957) 'Augenbefunde bei Frühgeburten.' *Klin. Mbl. Augenheilk.*, 131, 326.

Tranquillizers for Hyperactive Children

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It is better to use drugs than physical restraint to deal with the hyperactive child, but both should be a last resort. It has been our experience at this hospital that children who have been completely unmanageable at home, described as restless, never still, destructive and dangerous, often settle quite well in the ward. Modification of the environment and the routine of management should be given a careful trial before it is assumed that drug therapy is necessary. In an emergency, sedatives may be necessary to tide matters over until changes in the environment can be effected. It should always be remembered, however, that drugs may sometimes influence a difficult child's behaviour for the worse; this applies both to the longer established sedatives and to the 'tranquillizers' or ataractics. Barbour *et al.* (1936) drew attention to this problem in regard to bromides in psychotic and neurotic patients and it has since been generally recognised that disordered and overactive behaviour may be drug-induced.

Hyperactivity in regard to children is a blanket term covering several categories of disorder. A very backward child with an obvious physical basis for his disorder, such as tuberous sclerosis, is commonly overactive, largely because his limited intelligence prevents him from appreciating social situations and developing the normal inhibitions which regulate a child's behaviour in the presence of adults. At the same time he may not understand language, so that verbal admonition is

ineffective. Placement in a group, such as an occupation centre, by day, an opportunity for adequate physical exercise, and a definite routine will often adequately control behaviour in such cases and tranquillizers are likely to be indicated only occasionally.

The isolated deaf child is also not infrequently overactive. Here again drug therapy is not likely to be of great value. What is needed is individual attention and patient efforts to establish contact from an early age, whether by means of a hearing-aid, if acceptable, by attempts at teaching lip-reading, or by means of simple games in which meaningful contact can be made. Once contact is established, it becomes increasingly possible to control behaviour.

The most difficult group, and the one in which tranquillizers are worth a trial, are the children whose behaviour is quite inappropriate. This group includes both children of presumed normal intelligence and others who appear markedly backward. They display in some cases all the features of adult schizophrenia, modified by the patient's age and experience, while occasionally the child more resembles a case of hypomania. Many child schizophrenics have a greatly reduced output of energy and are hypoactive rather than hyperactive. In some cases, however there is continuous restless activity. In other children, particular symptoms may be a problem—continually running away, biting other children, head-banging and other forms of self injury, or obsessive-

compulsive activity in regard to beads or buttons, so that necklaces are ripped from unoffending necks and a whole cloak-room is denuded of buttons. It is this type of symptom which is most likely to arouse the annoyance of neighbours at home and be a difficulty for other patients in hospital.

There have been a number of enthusiastic reports of the value of tranquillizers in hyperactive children, of which the paper by Bair and Herrold (1955) is typical. When controlled trials are carried out, as has been done by Craft, the results appear less clear cut and convincing. His studies covered both adults and children. He found, among other things (Craft 1958), that meprobamate ('Equanil', 'Mepavlon', 'Miltown') appeared to have no useful effect on the patients' activity, even in the high dosage used (800 to 4800 mg. over three weeks) but that the direction of the activity might be changed. Thus, patients formerly attacking others now attacked themselves. In the case of hydroxyzine ('Atarax') (Craft 1957), no improvement was noted on a dose of 500 mg. daily, except in one ward which reported a highly significant change for the better both on the drug and on the placebo. Craft concluded his review with the statement that prochlorperazine was the best drug for the treatment of hyperactivity syndromes. He stressed, however, that his studies showed how important it was to recognise the importance of environmental change. He hoped that interest in the use of drugs might give added impetus to the abolition of physical restraint. The dose of prochlorperazine ('Stemetil'; similar to 'Largactil' and 'Thorazine') used rose to 150 mg. daily in the middle of a three-week trial. There was an overall reduction of behaviour on the activity scale, and only one patient showed undue sedation. The proportion of toxic reactions was, however, high, 4 out of 18 patients

showing parkinsonism, which soon disappeared on reduction of the dosage.

Trifluoperazine ('Stelazine') is in occasional use in this hospital for hyperactive patients and from time to time appears to produce useful results. Rettig *et al.* (1959) and Lawlis (1959) prefer it to prochlorperazine, though their studies were not controlled. The doses used by the first group were surprisingly small—1 mg. three times a day. Lawlis used an average of 6 mg. daily for boys of 7-15 years with no serious side-effects. We have found that for very severely disturbed patients of this age-range much larger doses are needed to produce an effect—e.g., 5 mg. four times a day. We have used thioridazine ('Melleril') also with apparently beneficial effects in some cases, in doses of 25 or 50 mg. three times a day.

The dosage of these drugs will need to be modified according to the children's age and weight, but it is difficult to lay down a standard since there seems to be a high degree of individuality in regard to both effectiveness and toxicity, so that a trial should be begun with a small dose which is increased gradually and reduced if toxic features appear. If behaviour improves on a drug, it should not be continued indefinitely until it has been established whether the improvement can be maintained without the drug. The dose should be reduced gradually and in many cases a placebo may be introduced with advantage, without the knowledge of the staff recording behaviour or of the parents, if the child is at home. A further course of the drug may be used to deal with a relapse in behaviour or to tide over a difficult situation. If social relationships are better while on a drug, the opportunity may be taken to introduce the child into a class or occupation centre, the dose being gradually reduced subsequently to see whether the pattern of group behaviour

once established can be maintained without the drug. Owing to the great individual variation in response to this group of preparations it is worth trying several if those first tried are not effective.

In conclusion, it should be emphasised again that drugs are at best an adjunct to the influence of the environment and personal relations. A hyperactive boy of 12 years recently broke a window, sustain-

ing deep lacerations of the arm. He was admitted to the casualty department of a general hospital where he caused consternation. Advice was requested as to sedatives and permission was being requested for a general anaesthetic. Meanwhile a male nurse who knew him well arrived on the scene, whereupon the boy meekly submitted to the insertion of a large number of stitches into his arm.

SUMMARY

The child who is hyperactive may be so for a variety of reasons. The condition does not constitute a syndrome. It is often possible to manipulate the environment and so avoid recourse to drugs which are, however, better than physical restraint. Drugs may be used as a temporary measure in a crisis or to initiate a change in the organisation of the child's life. Of the tranquillizers, prochlorperazine and trifluoperazine may be useful; in patients who fail to respond, other preparations such as thioridazine may be used. Dosage and choice of preparation vary for different children. Long-continued administration should not be permitted without testing the effect of gradual withdrawal.

RÉSUMÉ

Tranquillisants pour enfants hyperactifs

L'enfant hyperactif peut l'être pour diverses raisons. Cet état ne constitue pas un syndrome. Il est souvent possible d'agir sur le milieu ambiant et d'éviter ainsi de recourir à des substances médicamenteuses cependant meilleures que la contrainte physique. Les substances médicamenteuses peuvent être employées comme mesure temporaire au cours d'une crise ou pour introduire un changement dans la vie de l'enfant. Parmi les tranquillisants, la prochlorpérazine et la trifluopérazine peuvent être utiles. Pour les malades qui ne réagiraient pas, d'autres préparations telles que la thioridazine peuvent être utilisées. La posologie et le choix de la préparation diffèrent selon les enfants. On ne saurait en autoriser l'administration très prolongée sans contrôler l'effet de sa suppression progressive.

ZUSAMMENFASSUNG

Ataraktische Mittel für hyperaktive Kinder

Das hyperaktive Kind kann aus vielen Gründen so sein. Es ist oft möglich, auf die Umgebung zu wirken und so zu vermeiden, zu Drogen Zuflucht zu nehmen, die jedoch besser als körperlicher Zwang sind. Die Arzneimittel können als vorübergehende Massnahme für eine Krise oder um eine Veränderung in der Lebensorganisation des Kindes einzuleiten, angewendet werden. Unter den ataraktischen Mitteln können Prochlorperazin und Trifluoperazin nützlich sein; bei Patienten die nicht reagieren, können andere Präparate, wie Thioridazin verwendet werden. Dosierung und Wahl des Präparates sind verschieden für die verschiedenen Kinder. Langdauernde Verabreichung dürfte nicht erlaubt sein ohne Kontrolle der Wirkung der graduellen Entziehung.

REFERENCES

- Barbour, R. F., Pilkington, F., Sargent, W. (1936) 'Bromide intoxication' *Brit. med. J.*, *ii*, 957.
Blair, B. V., Herold, W. (1955) 'Efficacy of chlorpromazine in hyperactive mentally retarded children'. *Arch. Neurol. Psychiat. (Chicago)*, *74*, 363.
Craft, M. J. (1957) 'Tranquillizers in mental deficiency: hydroxyzine'. *J. ment. Sci.*, *103*, 855.
— (1958) 'Tranquillizers in mental deficiency: chlorpromazine'. *J. ment. Defic. Res.*, *1*, 91.
Lawlis, M. G. (1959) 'A note on trifluoperazine in the management of hyperactive mentally retarded children'. *In Trifluoperazine*. Edited by H. Brill. London: Kimpton, p. 180.
Rettig, J. H., Caldwell, W. L., Josephs, M. C. (1959) 'A pilot study of trifluoperazine in mentally retarded patients'. *In Trifluoperazine*. Edited by H. Brill. London: Kimpton, p. 173.

Man's Pilgrimage

First, birth:
The beginning:
The start of the way.

Then infancy:
An easy time,
Without a burden on your back.

Then childhood:
A happy road of flowers,
When all is gay and careless.

Then youth:
When you hope and dream and love.

Then maturity:
When cares set in,
And you may lose your joy of God
For love of earthly things.

Then age:
The end of earthly bondage.
And the sound of Trumpets you discern
For the wanderer's Return.

W. M. WELSH
(Age 12 years)

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Auto-antibodies and the Nervous System

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IMMUNE reactions of various kinds provide a good defence mechanism against infections. It seems a small step from these reactions of infection to the exaggerated immune response that is provoked by usually harmless dusts or pollen in an allergic illness such as asthma, although in this case the immune reaction is to the patient's disadvantage. In other diseases the immune response may be even more abnormal. Maternal antibodies against the Rhesus factor appear to be responsible for haemolytic disease of the newborn and so for many cases of kernicterus and athetoid cerebral palsy, while, in auto-allergic or auto-immune reactions, antibodies produced within the body are directed against the body's own components.

Ten years ago, most physicians considered that auto-immune diseases were freak disorders of laboratory animals that had no parallel in human disease. It was known that animals could be sensitised to their own tissues by the injection of tissue extracts from the same species. For example, injection of thyroid extracts could cause disease of the animal's own thyroid gland; injection of a homogenate of testis could cause testicular atrophy; and injection of brain emulsion could cause demyelinating disease of the recipient's own brain. The close resemblance of some of these disorders to well-recognised diseases of man led to the gradual realisation that auto-immune disease was not, after all, a mere freak of the laboratory. From Hashimoto's disease of the human

thyroid to the encephalomyelitis which sometimes follows vaccination against rabies, there is a whole range of human diseases that are now firmly established as disorders of the body's reactions, caused by a hypersensitive or abnormal immune response.

When Billingham, Brent and Medawar (1953) studied immune reactions in the foetus, they found that at this stage of life there was complete tolerance to all kinds of foreign materials which might be introduced into the body from outside. They showed, moreover, that once a foetus has been in contact with a foreign substance, its tolerance to it will persist even when the animal is more mature. Thus anything present in the body or encountered by it up to a certain stage of development seems to be recognised as part of itself, anything introduced later being treated as a foreign substance which then provokes immune reactions of various kinds. It follows that complete tolerance towards the various components of the body is normal, and it is probably only in three circumstances that this tolerance is lost: first, if a substance that is normally segregated is accidentally released into the blood-stream and thereby comes into contact with antibody-producing cells; secondly, if a normal component of the body is so altered by disease, or by drugs, or experimentally by 'adjuvants', that it is treated as a foreign substance by the antibody-producing cells; and lastly, when there is a disorder of the reticulo-endo-

thelial system itself, which may become overactive and over-responsive to antigenic stimuli.

Diseases from Disordered Reactions

Some of the diseases caused in these ways are now well recognised. When, during an operation, a surgeon releases protein from the seclusion of the lens of the eye, antibody to this protein may lead to sympathetic ophthalmia in the opposite eye. Similarly, the liberation of thyroglobulin, which does not normally come into contact with the circulation, may act as the first stimulus in the development of a chronic thyroiditis. When the drug 'Sedormid' is given to certain subjects, it may cause such changes in the blood platelets that they are treated by the body as foreign material. Antibodies to the altered platelets lead to their destruction, and purpura results (Ackroyd 1949). An abnormality or overactivity of the reticulo-endothelial system itself may also cause disease and is probably present in Hashimoto's thyroiditis and in systemic lupus. In these diseases, not only is there a tendency for the patient to produce a number of different auto-antibodies, but antibodies may also be found in apparently healthy members of the same family (Hall, Owen and Smart 1960; Pollak, Mandema and Kark 1960).

Theoretically, the onset of an immunological disease might be expected to follow at a predictable interval after an infection or antigenic stimulus. It might also be expected that the disease would be associated with lymphocytic infiltration, with globulin deposits in the affected tissues, and with the release of antibody globulin into the circulation. Steroids should be beneficial, and finally, the injection of the appropriate antigenic substance should produce a similar condition in animals.

It is perhaps apt, in this context, to

consider a neurological disease which has many of these features—namely, the demyelinating encephalomyelitis which may follow vaccination against rabies. The vaccine which provokes the disease is made of rabbit brain or spinal cord, and the subsequent reaction develops too late to be accounted for by an inflammatory reaction to the injected material. There is also strong circumstantial evidence for the auto-immune origin of this reaction since, histologically, it bears an extremely close resemblance to the allergic encephalomyelitis of animals caused by injecting myelin-containing brain extracts together with Freund's adjuvant. As yet, this rather rare condition is the only nervous disorder in the human subject in which the evidence for an auto-immune origin is widely accepted, though a similar cause is probable in postinfective encephalomyelitis and postinfective neuritis (Miller, Stanton and Gibbons 1956). In support of this, postinfective neuritis strongly resembles the allergic polyneuritis which has been produced in animals (Waksman and Adams 1955).

Multiple sclerosis and other demyelinating diseases of childhood and adult life also have some features which might suggest an auto-immune disease (Kolb 1950). Multiple sclerosis is often associated with a raised gamma-globulin level in the cerebrospinal fluid (Colover 1961) but not with a raised globulin level in the blood. In this it resembles allergic encephalomyelitis of monkeys (Allegranza 1959), though the histological resemblance is less striking than in the case of post-vaccinal encephalomyelitis. Like the experimental disease, multiple sclerosis responds rather poorly to treatment with steroids, and exacerbations cannot be prevented by this means. The serum of many patients has been shown by Frick (1954) to contain complement-fixing antibodies to a lipoprotein extract of normal brain. However,

despite this evidence, it is not established that multiple sclerosis is an auto-immune disease, and further research is needed to exclude the possibility that the disease may, after all, be a virus infection, in which the antibodies that have been detected are the secondary results of an infective process.

Future Studies

In human diseases which have the clinical features of an immune disorder and which resemble allergic disorders of laboratory animals, a great deal is still likely to be learnt as the result of immunological studies of the serum proteins, of skin hypersensitivity, and of globulin deposits in the affected tissues. Diseases of the nervous system offer a stimulating field for work of this kind, but it cannot be assumed that the presence of auto-antibodies means they are the cause of a disease. In every known disorder of the thyroid gland in which there is damage to thyroid tissue there have been reports of the presence of antibodies to thyroglobulin, presumably arising as the result of a secondary reaction to tissue breakdown. This cannot mean that all these diseases

are caused by antibodies, though in auto-immune thyroiditis, where similar antibodies are present in extremely high concentration, they almost certainly play an important part in causing the disease. In auto-immune thyroiditis and other thyroid disorders there is probably a distinction between antibodies which have a damaging effect and those which arise as part of a secondary immune response (Lesso, Crawford and Wood 1959). There may be a similar need in disease of the nervous system to distinguish between harmful immune reactions and those of a secondary nature. As immunological techniques improve, an increasing number of diseases are found to be associated with hypersensitive reactions and antibody production; but even in cases in which hypersensitivity to nervous tissue can be demonstrated, or in which there are tissue deposits of globulin, the final key to the disease may lie in knowing whether these reactions are harmful or merely secondary; what is the nature of the trigger mechanisms that fire off the response; and whether the antibody-forming cells are primarily deranged. Studies of this kind are still in their infancy.

SUMMARY

In auto-immune disease, antibodies are produced within the body which react with the body's own components. Experimentally, the injection of tissue extracts in animals may result in thyroiditis, encephalomyelitis, or other diseases; while, in man, sympathetic ophthalmia, Hashimoto's thyroiditis, allergic purpura, and various types of demyelinating disease have all been suspected of having an auto-immune origin. However, the presence of circulating antibodies which react with appropriate tissue extracts is not in itself evidence that they are the cause of disease, for these antibodies may arise as a secondary reaction to tissue breakdown.

RÉSUMÉ

Les auto-anticorps et le système nerveux

Dans l'auto-immunisation, les anticorps sont produits dans l'organisme qui réagit avec ses propres composants. Sur le plan expérimental, l'injection tissulaire chez l'animal peut aboutir à une thyroïdite, une encéphalomyélite ou à d'autres affections. Chez l'homme on soupçonne l'ophtalmie sympathique, la thyroïdite d'Hashimoto, le purpura allergique et

divers types de myélite d'avoir une origine auto-immune. Cependant, la présence d'anticorps circulants réagissant aux extraits tissulaires appropriés n'apporte pas en elle-même la preuve qu'ils sont à l'origine de la maladie; ces anticorps peuvent fort bien n'être qu'une réaction secondaire à la détérioration des tissus.

ZUSAMMENFASSUNG

Die Auto-Antikörper und das Nervensystem

Bei Auto-Immunisierung bilden sich die Antikörper im Organismus, der mit seinen eigenen Komponenten reagiert. Experimental kann Injektion von Gewebsextrakten bei Tieren zu Thyroiditis, Encephalomyelitis oder anderen Krankheiten führen. Beim Menschen hat man vermutet, dass sympathetische Ophthalmia, Thyroiditis, Von Hashimoto, allergisches Purpura und verschiedene Myelitisformen einen auto-immunen Ursprung haben. Die Anwesenheit von zirkulierenden Antikörpern, die mit entsprechenden Gewebsextrakten reagieren, ist in sich jedoch kein Beweis dafür, dass sie die Ursache der Krankheit darstellen, denn diese Antikörper können als eine sekundäre Reaktion auf Gewebsschädigung entstehen.

REFERENCES

- Ackroyd, J. F. (1949a) 'A simple method of estimating clot retraction, with a survey of normal values and the changes that occur with menstruation.' *Clin. Sci.*, **7**, 231.
 — (1949b) 'The mechanism of the reduction of clot retraction by sedormid in the blood of patients who have recovered from sedormid purpura.' *Ibid.*, **8**, 235.
 Allegranza, A. (1959) *In Allergic Encephalomyelitis*. Edited by M. W. Kies and E. C. Alvord, Jr. Springfield, Ill.; C. C. Thomas; p. 490.
 Billingham, R. E., Brent, L., Medawar, P. B. (1953) 'Actively acquired tolerance of foreign cells.' *Nature*, **172**, 603.
 Colover, J. (1961) *Proc. Ass. clin. Path.* (In the press).
 Crawford, H. J., Wood, R. M., Lessof, M. H. (1959) 'Detection of antibodies by fluorescent-spot technique.' *Lancet*, **ii**, 1173.
 Frick, E. (1954) 'Zur Serologie der multiplen Sklerose.' *Klin. Wschr.*, **32**, 450.
 Hall, R., Owen, S. G., Smart, G. A. (1960) 'Evidence for genetic predisposition to formation of thyroid autoantibodies.' *Lancet*, **ii**, 187.
 Kolb, L. C. (1950) 'The relationship of the demyelinating diseases to allergic encephalomyelitis.' *Medicine*, **29**, 99.
 Miller, H. G., Stanton, J. B., Gibbons, J. L. (1956) 'Para-infectious encephalomyelitis and related syndromes.' *Quart. J. Med.*, **25**, 427.
 Pollak, V. E., Mandema, R., Kark, R. M. (1960) 'Antinuclear factors in the serum of relatives of patients with systemic lupus erythematosus.' *Lancet*, **ii**, 1061.
 Waksman, B. H., Adams, R. D. (1955) 'Allergic neuritis: an experimental disease of rabbits induced by the injection of peripheral nervous tissue and adjuvants.' *J. exp. Med.*, **102**, 213.

REPORT

Ontogenesis of the Central Nervous System

PILSEN SYMPOSIUM: SEPTEMBER 1960

Reported by DR. ALEX MINKOWSKI

At the beginning of September, 1960, a symposium sponsored by the Purkinje Society was held in Pilsen (Czechoslovakia) on the Ontogenesis of the Central Nervous System. It took place in the public library in an old convent where the Czech composer Smetana once lived.

Important experimental work was reported from various institutes in the USSR, all specialising in development neurophysiology, and from two institutes in Czechoslovakia (*Pilsen: Dr. Mysliveček; Prague: Dr. Sedláček*). Most of this work was concerned with conditioned reflexes in the animal and also in man.

Some of the research (*Sedláček*) was based on the generalisation of **A. A. Volokhov** concerning developmental nervous activity. It was shown that in the rat, development of reflex action begins with the simplest forms of reactions of the organism to stimulation of the skin—with local reflexes acting through the first elementary synoptic connections in the spinal cord (15th–16th day to 19th day of gestation). Following the period of local reflexes, a generalised reaction mediated by the whole spinal cord is typical after the maturation of ascending and descending pathways (20th day of gestation to birth). With the maturation of morphological structures in the medulla oblongata, the phase of primary generalisation of response is replaced by slow generalised tonic reactions requiring activity in the proprioceptive afferents (1st to 6th day after birth). The phase of specialised reflexes (washing, shaking, palpebral and

other reflexes) begins with the maturation of the midbrain (7th–8th up to 14th–17th day). This concludes the first stage of the development of reflex activity based on the emergence of inborn unconditioned reflex mechanisms. The second stage of development of the conditioned reflex mechanisms begins with the phase of inhibition of skin specialised reflexes, which is connected with the development of the higher levels of the central nervous system above the midbrain (14th–17th up to the 20th day). The next developmental stage is characterised by 'spontaneous' specialised reflexes in the form of simple conditioned motor reflexes (15th–25th day). The last phase of development of reflex activity is the elaboration of complex 'spontaneous', specialised, conditioned motor reflexes (starting from 25th day). Therefore the development of reflex activity in the ontogenesis of the rat follows the principle of development in stages, whose bases are the qualitative changes of function and structure of the central nervous system.

A. A. Volokhov illustrated this theory by a film on the development of nervous activity in animals.

In a morphological study on the co-operation of analysers in conditioned reflex activity, **O. S. Adriyanov** (*Institute of the Brain, Academy of Medical Sciences, Moscow*) found no support for the correlative and integrative rôle of the so-called non-specific (reticular) formation of the diencephalon and mesencephalon in the forming of conditioned reflexes and

in the process of co-operation of analysers.

B. N. Klossovski (*Institute of Paediatrics, Academy of Sciences, Moscow*) thought that brain structure in ontogenesis was influenced by two factors in its development, the basic material and external stimuli. This last idea was supported by various experiments in which both caudate nuclei, both thalami, visual and motor-analysers and colliculi superiores were removed. According to **Klossovski**, impulses from the external environment stimulate maturation of the nerve cells, myelinisation of their network, and angio-architecture in the brain, alongside the pathways of impulses from the periphery.

A. B. Kogan's (*Rostov on Don*) experiments provoked some controversy. He stated that if visual or acoustic cortical analysers were removed at an early age, visual and acoustic analysis was no worse and might be better in these animals than in controls, although some defects of the nervous tissue were evident after one year.

J. Mislivěček (*Institute of Pathophysiology, Charles University, Pilsen*) showed, on the other hand, that removal of the neopallium in young albino rats caused regression in development resulting from deterioration of the highest regulatory functions and probably also from the persisting changes in the dynamics of intermediary metabolism. The experiments that he demonstrated on the conditioning of rats were so consistent that **Mrs. Flexner** proposed that the rats should be awarded a medal.

A. I. Karamyan (*I.M. Setchenov Institute of Evolutional Physiology, Academy of Sciences, Leningrad*), studying fishes, found that all afferent and efferent pathways met in the visual regions of the midbrain and cerebellum.

Another topic was the correlation between electrophysiological findings and

biochemical and morphological changes during ontogenesis. This was mainly studied by **L. B. Flexner** (*University of Pennsylvania, Philadelphia*), working especially on lactic dehydrogenase, by **Z. D. Pigareva** (*Institute of Normal and Pathological Physiology, Academy of Medical Sciences, Moscow*), who together with the Russian neurobiologist Krebs, studied succinodehydrase, cytochromoxidase and cytochrome systems in the tissue of various regions of the central nervous system, and by **Mislivěček**, working along the same lines.

It must be stressed that, in its present stage, this sort of work, although promising, should be restricted to coincidences in ontogenesis rather than demonstrated correlations between biochemical changes, histological maturation and bioelectrical activities.

In other research, at the *Centre de Recherches Neurophysiologiques de l'Association Claude Bernard, Salpêtrière, Paris*, Marty and Contamin studied electrocortical responses to visual stimulation in newborn rabbits and cats. Working in the same Institute along similar lines, **J. Scherrer** and **R. Verley** have compared spontaneous and provoked activity (local as evoked potentials and general). This is in contrast with almost all the work already described on conditioned activity.

This brings some support to Coghill's theory of prenatal development of neurogenic movements. **H. Tuge** (*Hosei University, Tokyo, Japan*), a former co-worker with Coghill, has developed the theory that, in embryos of fishes, amphibia and reptilia, the earliest neurogenic somatic movement is a total response of head and trunk, and later the movement of appendages participates in the total response. Following these responses the discrete and localised reflex of appendages becomes evident. In bird embryos, although opinions are divided, it may be concluded that

the generalised reflex activity of the whole body appears first, the localised reflexes following. Accordingly, the development of somatic behaviour in the prenatal period in inframammalian vertebrates appears to abide by the Coghillian concept.

There are two schools which object to the Coghillian concept of prenatal development of somatic function in mammals, including man—namely, Volokhov's and Windles'. Alex Minkowski (*Clinique Obstétricale Baudelocque, Paris*) examined their findings from the point of view of the methods employed, the morphogenesis of the nervous system involved, in connection with the development of reflex activity, and theoretical considerations, particularly on the evolutionary basis. The view that the reflexes of total pattern precede those of local pattern during embryogenesis, even in mammals, seems to be substantiated. It is suggested that in order to elaborate a localised reflex activity, the ontogenetic development of central inhibitory processes is, from the physiological point of view, indispensable at the time when an individual reflex arc emerges and becomes differentiated morphologically in the central nervous architecture. In the postnatal period, expansion of the adaptive activities necessitates the development of conditioned reflex activities on the basis of unconditioned reflexes. In studying the ontogenetic development of conditioned reflexes from the lower to the higher animals, particular attention should be paid to the genesis of the orientation reflex, which shows that the generalised response becomes localised and refined as development proceeds. Various kinds of conditioned reflexes in their ontogeny begin with a diffused generalised type and later become localised and discrete. All the facts so far observed appear to indicate that the nervous function of unconditioned and conditioned reflexes throughout the pre- and post-natal periods of animals develops at first in a diffused

and generalised form and later acquires a localised and refined form. This concept seems to be one of the fundamental principles in nervous activity, when viewed from a correlated study of structure and function.

Only one paper was given on ontogenetic anatomy, by E. Horne Craigie (*University of Toronto, Canada*), who dealt with vascularity in the visual centres of the rat's brain.

A limited part of the symposium was devoted to human ontogenesis. Alex Minkowski reported S. Saint-Anne Dargassies' and C. Dreyfus-Brisac's work (*Centre de Recherches Biologiques Néonatales de l'Association Claude Bernard, Clinique Obstétricale Baudelocque, Paris*). Parallel development of electroencephalogram patterns and spontaneous activity have been observed in 21 human foetuses aged 4 and 5 months. The developmental neurology of the premature infant was illustrated in Mme Saint-Anne Dargassies' film, which gave rise to an interesting discussion by I. A. Arshavski.

The Pavlovian School investigated the ontogenesis of conditioned reflexes. H. Papoušek (*Institute for Maternal Care, Prague*) studied the voluntary activity of infants who received sweetened or unsweetened milk, according to which side they turned their head at a given signal; the child gradually begins to turn only to the side of the sweetened milk and seeks it on the other side when the experimental situation is reversed.

N. Y. Kasatkin (*Setchenov Institute of Evolutional Physiology, Academy of Sciences, Leningrad*) gave a general review of the development of conditioned reflexes. The first conditioned reflexes which arise in human ontogenesis are the main criterion of the earliest activity of the higher brain centres in man. In the course of individual development early conditioned

reflexes are formed in the postnatal period. Intrauterine conditioning in the child has not yet been proved. The elaboration of artificial conditioned reflexes in premature children shows that it is possible to develop such a reflex after delivery before the normal full-term is reached. In the first few weeks of life, normal healthy children form conditioned reflexes in a definite order. Conditioned reflexes are first formed within the limits of the vestibular and auditory analysers, and later within the limits of the tactile and visual analysers. A relationship is postulated between the appearance of the earliest conditioned reflexes and the development of rhythm of some physiological processes shortly after birth. The time-conditioned reflexes of humoral changes, or the changes of total motor activity in the newborn, can be taken as examples. In contrast to the adult organism, the

formation of the first conditioned reflexes in children is predetermined by the morphological maturity and degree of functional readiness, first, of the central structures of the brain; secondly, of the peripheral receptor apparatus; and thirdly, of the effector or reacting apparatus.

On the whole, this was a fruitful meeting. There is still some gap between the eastern and western schools of neurophysiology, the eastern school being mainly interested in conditioned reflexes and the western in spontaneous or evoked bioelectric activity. Both sides undoubtedly gained, not only from the scientific standpoint but also from that of international friendship which prevailed at Pilsen, thanks to **Dr. Mysliveček** and **Dr. Sedláček**, the main organisers.

LETTERS TO THE EDITOR

Minimal Cerebral Palsy

SIR—I was interested to read Dr. Wigglesworth's letter on this subject in the June *Cerebral Palsy Bulletin* (1961, 3, 293-295). I am sure he is right in drawing attention to this syndrome, which has often gone unrecognised in the past. Professor Donald Court, Dr. Errington Ellis and I have recently studied a series of children who have attended the Royal Victoria Infirmary, Newcastle-upon-Tyne, and the Percy Hedley School for Spastic Children. Each of these children had at some stage or other been regarded as mentally backward because of serious disorders of motor activity and of educational difficulties which resulted from their constitutional clumsiness. In each of the five cases which we have studied in detail there was no evidence of any spasticity or of weakness or ataxia of the limbs. On studying these children carefully, however, it became apparent that their difficulties in learning, in both the physical and the intellectual sense, were related to developmental defects of an apraxic or agnosic character. These defects only came to light when these children were studied carefully, and it was found that they showed certain specific defects of skilled motor activity or of visual or sensory recognition involving such activities as dressing and undressing, and also writing, copying and drawing. In several of these patients the disability resulting from these defects was so severe as to render the child almost ineducable in an ordinary school, and it was easy to see why in every case a diagnosis of mental backwardness had been considered at some stage by competent medical observers or by experienced school-teachers. In fact, estimation of the I.Q. in these cases, using the Wechsler scale for children, revealed in each one a striking discrepancy between a high or normal verbal reading and a greatly reduced reading on the performance scale.

We have concluded that this syndrome of congenital apraxia or agnosia is not uncommon in a severe form, and probably exists in a mild form in many children who are simply regarded as being constitutionally clumsy or difficult to educate. We feel that in all probability this syndrome is due to a disorder of cerebral organisation akin, in certain respects, to that responsible for congenital reading defects. We think that this explanation more probably accounts for the condition than would a focal cerebral pathological change resulting from hypoxia or birth injury. This view is supported by the total absence of other neurological abnormalities in the majority of these cases and by the fact that many of the patients are left-handed or ambidextrous, and it may be assumed that hemisphere dominance has not become properly established in one or other cerebral hemisphere. It is important for this condition to be brought to the attention not only of paediatricians, neurologists and family doctors, but also to school-teachers who may thereby be encouraged to recognise the special needs of children with this kind of disabling defect at a very much

earlier stage. In our experience, patient individual training pays considerable dividends in these patients, and it is clearly important that they should be recognised as early as possible.—Yours, etc.

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Newcastle-upon-Tyne, 1.

JOHN N. WALTON

Speech Therapy for Developmental Disorders

SIR—Miss Renfrew is to be congratulated on her attempts to assess the results of her therapy on children with developmental speech disorders. I would like to learn more about the tests that she uses than she gives in her letter and know the actual proportion of patients that she finds derive benefit. In particular I wonder if she really uses 'standardised' tests (i.e., tests validated by experience of their use in a large random population) or merely 'standard' tests. Perhaps she might be persuaded to write an article for the *Cerebral Palsy Bulletin* about her work.

I agree with her that I should have included in my *Bulletin* article some details of the patients I examined and the methods of examination used. Some, though unfortunately not all, of the details about my patients and methods which she requires can be found in an article in *Brain*, 1959, 82, 450, a reprint of which I sent her on December 10, 1959.

The majority of the patients were over the age of four years but had not reached the age of six at the time of their first examination. Most of them were seen at three-monthly intervals, but a few, especially those coming from great distances to Edinburgh, were seen only six-monthly. As far as possible, the children who received treatment mostly from Edinburgh were matched with those who did not receive treatment, mostly from outwith Edinburgh, by age, sex, place in the family, opportunity for play with other children and severity of the retardation of speech development. The severity of speech defect was measured rather crudely by two criteria. (1) The numbers of consonant sounds which were constantly and inconstantly defective; and (2) the parents' statements as to whether the child was intelligible to other children only, intelligible to the mother as well as other children, intelligible to the father as well as the mother, intelligible to other adults who knew the child as well as the parents, or intelligible to all strangers, but only with difficulty. Progress was assessed at one year and again at two years if the children were still attending the clinic.

I would be the first to agree that these attempts at assessment are crude in the extreme, but at least I was not directly concerned with the patients' therapy and was neither biased in favour of finding therapy useful or useless. In fact Miss Renfrew's results and my own are very similar. Patients very often show a 'spurt' in speech development which is not expected in the course of spontaneous maturation within six or twelve weeks of beginning speech therapy, and these 'spurts' are unusual in untreated children. They are found, however, in some patients who are placed in the company of other children for the first time, having been deprived of it previously—for example, after they have been admitted to nurseries or play centres. These spurts are usually succeeded by a period of relatively slower speech development and most patients, treated or untreated, acquire consistently correct articulation of all consonants only at the age of between six and seven years, as Miss Renfrew states.

Occasionally, because a child is passing through a period of 'real frustration', speech

therapy may be indicated in an attempt to secure a spurt in speech development. But it must be recognised that the frustration is more often on the part of the parents than the child and that frustration may often be diminished as well by direct advice to the parents on the management of their child. Therapists often forget how much they ask of parent and patient in terms of effort, money and family stress when they arrange for regular clinic attendances. I think that children of six years or more with developmental speech defects more often benefit from attending a speech therapist than those under this age.

How much they benefit because they find an adult who is understanding of their disability and prepared to try to help them to overcome it, and how much is due to the direct application of the techniques of correcting articulatory patterns is very difficult to determine.

The disagreement between Miss Renfrew and myself really comes down to one of opinion, for I do not dispute her facts and I admire her rhetoric. She believes that it is worth while treating retarded speech development by speech therapy because some children show accelerated speech development for a period as a result. I do not think that the spurt occurs frequently enough or is sufficiently important to merit regular attendance at a speech therapy clinic in most cases, especially in pre-school children. Further, it is often possible to achieve a similar spurt by placing a child in closer contact with other children than he has previously experienced. Both the parents' anxiety about the speech defect and the child's (which is usually secondary) can be greatly relieved by careful discussion of the cause and the management of the speech disorder. This is often the only treatment required.—Yours, etc.

Department of Child Life and Health
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T. T. S. INGRAM,
Lecturer

Maternal Diabetes and Cerebral Palsy

SIR—In the course of social work among cerebral palsied children I observed that a large percentage of these children had diabetic mothers. Most diabetic women in the child-bearing age are severely affected and require treatment with insulin, and it would seem possible that the occurrence of cerebral palsy in their offspring is related to the hypoglycaemia caused by this.

It is a recognised fact that women suffering from diabetes are more likely to bear prematurely, and that premature babies have a somewhat higher rate of handicapped conditions, such as cerebral palsy, mental retardation, epilepsy and physical malformation, than those born at term. The proportion of cerebral palsied children among premature babies is ten times as great as among babies delivered at full term.

I have known 8 cases of cerebral palsy and 1 of epilepsy born to diabetic women (about 20 per cent of those under my observation), and in a further 6 cases the mothers were obese and may well have had a prediabetic type of sugar curve.

It would be an interesting line of research to find out whether women who have given birth to cerebral palsied children are specially liable to hypoglycaemia caused by starvation, or whether they show an increased insulin sensitivity.—Yours, etc.

Richmond,
Surrey.

MAUREEN HOLMES MAWHINNEY

Miss Mawhinney's letter has been shown to Dr. Alison D. McDonald, of the Paediatric Research Unit, Guy's Hospital, who replies as follows:

The high foetal death-rate in maternal diabetes is well known, and there is some

suggestion of a raised incidence of malformations among the children of diabetic mothers. The only published reference to cerebral palsy that I have found is in the survey by Asher and Schonell¹. In 3 of 349 cases of cerebral palsy the mothers gave a history of diabetes, which was higher than the incidence of diabetes in the patients at a maternity hospital; but the criteria used may well have differed. In another survey in which mothers were asked about diabetes, there was only one instance of maternal diabetes among 240 cases of cerebral palsy². A slightly raised incidence of cerebral palsy may, as Miss Mawhinney has observed, be explained by a tendency to premature delivery of diabetic mothers. Further enquiry about the cases Miss Mawhinney mentions might well be profitable. Certainly further evidence on the possible connection between the two conditions is desirable before any investigation of hypoglycaemia or insulin sensitivity is undertaken.

Alison D. McDonald

¹ Asher, P. and Schonell, F. E. (1950) 'A survey of 400 cases of cerebral palsy in childhood.' *Arch. Dis. Child.*, 25, 360.

² Mitchell, R. G. (1961) Personal communication.

NOTICE

CATALOGUE OF SELECTED FILMS ON PEDIATRICS AND CHILD HEALTH

THIS Catalogue¹ is the outcome of a long-term project by the Committee on Medical Education of the American Academy of Pediatrics and the Department of Medical Motion Pictures and Television of the American Medical Association. Only films selected by the reviewing committees have been included and these number some 130 titles, though only a minority are directly obtainable from the sponsoring bodies.

The Catalogue follows the tried pattern, starting with the film title, followed by technical information concerning size, colour, sound or silent, and running time in minutes. Full credits are given to the producers and authorities consulted, together with the distributors' address and loan conditions. Also, following the admirable American practice, information is given on the purchase price of prints as well as on the cost of rental. Where the film is said to be on loan this presumably means free loan.

We are given a paragraph on the film's content, together with observations on its technical qualities and treatment, as well as indications of the possible audiences that the film is suitable for, and in many cases, for which it is recommended. The vast majority of the films are of course American in origin, but the odd title or so crops up from other sources, such as Canada and Great Britain.

The Catalogue is divided into two parts: Films for Professional Audiences and Films for Lay Audiences. Within these classifications the films are broken down by broad subjects—Nutrition, Pharmacology, etc.—and by alphabetical order within the subgroup.

The whole work points to considerable industry and is hard to fault except on minor points, such as a better lay-out of the technical information so that important features are immediately recognised. One weakness of classification in combining the *Contents* with the *Index* is that no overall alphabetical index is available. Such small defects, however, do not detract from the value of this work, the first of its kind on the subject.

Frank Bamping

¹ Obtainable from the American Medical Association, 535, North Dearborn Street, Chicago 10, Illinois, or the American Academy of Pediatrics, 1801 Hinman Avenue, Evanston, Illinois, U.S.A. pp. 52, Gratis.

BOOK REVIEWS

The Doctor, his Patient and the Illness

By MICHAEL BALINT

London: Pitman Medical Publishing Co., 1957, pp. 356, 40s.

Reviewed by CYRILLE KOUPERNIK

This book is the result of a living and unusual collaboration. A great effort of mutual understanding and co-operation has been made, apparently for the first time in this particular way, between the members of two previously diverging medical trends, general practice and psychiatry. Such a divergence is unfortunate, and is not in accordance with factual reality, as is lucidly put forward in this book. For it must be obvious that the human material entrusted to the one or the other branch is the same, and only the techniques of approach and treatment differ. This common human material extends beyond what we label as the neuroses, and besides, this is a field in which nosology soon proves to be stifling and sterile. It is Man who is in question, Man in everyday life, Man with his psychological difficulties, which he is almost compelled to clothe in the garb of somatic illnesses, because it is to his general practitioner that he will appeal for help. This medical attendant enjoys his whole trust, they share a common way of life in the same locality, and this is certainly the main advantage that the practitioner has over the more distant and inaccessible specialist. If the situation is now considered as a whole, the practitioners score on another point: owing to their great numbers, and because they have presumably become used to proceeding rather rapidly, they will show a more efficient return than the qualified psychia-

trists. But the latter, who do not have the daily contact, and in spite of their lower practical efficiency, have, as a rule, a much better knowledge of the theories explaining the mental processes of their patients.

As Balint humorously remarks, the seminars of the Tavistock Clinic have made it clear that the drug most commonly prescribed by the doctor was himself, in other words his own psychological action. The meetings which gave rise to this book were held regularly at the Tavistock Clinic in order to investigate this apparently new situation. They consisted of a group of one or two psychiatrists and eight to ten general practitioners and were held every week for two to three years. This group teaching has become part of the way of life of anglo-saxon countries, in which the magisterial teaching which flourishes on the continent is regarded with some suspicion. In the present instance this situation is even more unusual: for its object is to study a first type of relationship, that which is established between the practitioner and the patient, but this investigation is carried out within the framework of another relationship, that which exists between the practitioner and the specialist. In general, Balint's whole effort is to warn the practitioner against an attitude which the physician is only too prone to adopt, that of the infallible schoolmaster. Indeed can one imagine a better way of teaching this passiveness, this humility, than that of practising it

oneself? In other words a new balance must be struck between the practitioner and his sometime teacher, the psychiatrist or qualified psychotherapist. And indeed this teaching is not a one way process inasmuch as the specialist becomes initiated in a daily life which would normally be totally outside his ken.

It is impossible to analyse this book in detail, although every line of it is worth reading, especially for the case-histories given by the practitioners. Such histories may be compared, more particularly, with the standard case-histories, summarised by a physician with an equally standard background, and which relate a number of allegedly objective data which are, in fact, completely devoid of medical, scientific or human interest. On the other hand, the histories of the 'dynamic' type related by those taking part in the seminar, gradually reveal, through the patient's symptomatology—this notorious functional symptomatology which shows no clear evidence of which function is involved—a new dimension which is that of the doctor-patient relationship. According to Balint, the deep significance of the symptoms resides in the offer which the patient makes to the practitioner; the latter may accept them or reject them. If, after a number of objective examinations and references to the specialist, the practitioner informs the patient, who still comes to see him, that there is no disease, this means that the practitioner refuses the patient's offer and declines to give the help which is implicitly sought. On the other hand, if he accepts the offer in the somatic form, he must make up his mind never to touch on the root problem and by so doing he will cause the patient's symptoms to endure. In other words there is only one solution for the practitioner, the least facile inasmuch as rejecting the offer and sheltering in organic pseudo-medicine constitutes an easy solution, the doctor must approach

his patient in a psycho-therapeutic spirit. Here then is how the practitioner's position appears as he faces what is, ultimately, his main task: alone he must deal with this problem which he is not trained to meet. Only those practitioners who have had the good fortune to attend the Tavistock Clinic seminars have received intelligent and adequate help. The others stand alone; it is not even advisable for them to call in a consultant, inasmuch as such a step dilutes their responsibility and gives rise to that which Balint calls the 'collusion of anonymity'. Balint even considers that there is no indication for proceeding with numerous complementary investigations having as their object the ruling out of organic disease masquerading as a neurosis. This aspect of the problem is indeed essential; Balint is right and he is also wrong. By that it is meant that if this immediate plunging into psychotherapy has definite advantages from the psychological point of view, it nevertheless creates a risk, for a limited but perhaps not a negligible number of cases, of the occurrence of alarming diagnostic errors. There is a general tendency to encourage practitioners to develop a facility for making emergency diagnoses; it seems inadvisable to give up this policy; if priority is given to the psychological features there is a grave danger of lowering the general standard of medical care.

Yet other dangers threaten the doctor and the patient in this new relationship. Even in the event of a psychological situation, psychotherapy may be misguided and result in a deterioration. There is a word which Balint hardly mentions, but for professional psychiatrists it is part and parcel of the process of watchfulness; the word is 'suicide'. The author is quite right when he teaches that psychotherapy is no mere patting of the patient's cheek or shoulder, nor a mere permanent encouragement, it should come to the

heart of the matter, it may have to cope with a matter of life or death. But under these conditions, the burden placed on the practitioner's shoulders may be too heavy, or else contact with the patient may be lost and he may drop to a lower level than that of his starting point. To make up for this however, the problem of the cessation of treatment is possibly better resolved when the practitioner administers it, as there is always the possibility of a persistent psychotherapeutic relationship beyond the period of active treatment.

The essential point of the book may now be considered again, that the physician prescribes himself in the same way as a drug. We must know what the taste of the drug is, and what its action is. These various aspects of the doctor's personality, and, in particular, of the part he feels he should play, are analysed with great clarity in two chapters entitled: 'the apostolic function of the physician.' Indeed it appears, to quote the author (p. 216): 'it was as if each doctor had experienced a kind of revelation concerning what is beneficial and what is harmful for the patients, and even more as if he had a sacred duty to convert to his own faith the ignorant and the wrongdoers among his patients'. Many other aspects of his personality at least as it is felt by himself and by his patients, lead to a reinforcement of this conviction of an apostolic role: the doctor visits the patients, he must bring them comfort, finally a doctor is a confessor, anything can be said to him, he will understand everything and repeat nothing. There remains a last aspect, which is not the least important, the doctor is most likely to react positively towards a patient who believes that he has improved under his care, and on the contrary, to interpret a failure to improve as a true sign of aggressiveness.

The next chapter may also be read with the greatest interest, it appeared to the

reviewer to be too short and is entitled 'the patient and his disease'. It must not be forgotten, incidentally, that these terms are actually part of the title of the book. In point of fact this chapter is not as inspired as the others insofar as it introduces theoretical concepts which are far from being authenticated. In it is shown one of the latest theories of psychosomatic medicine of analytical origin; i.e. that there is in each individual a kind of basic illness or defect in the biological structure, and that this is at the root of all future pathological changes. According to this view, the special function of the illness is that it is an insoluble conflict, insoluble either owing to too heavy a burden of exogenous factors, or to the involvement of a sort of Achilles' heel which is the basic defect. This theory is certainly very neat, but it seems somewhat artificial to unite in the same dynamic motion, as the author does, stillbirth, Huntington's chorea, an overwhelming infection or a bomb cast by the enemy. Likewise, for Balint, when the subject must face a problem which, for some reason or other, he cannot resolve, after a variable latent period he will develop one disease or another. But can one truly extend this explanation to all the conditions which are encountered? The author seems to think that there may be psychological reasons for the rare incidence of cancer under the age of 40, while on the other hand this age tends to mark the upper limit for ulcers; would it not be more logical to consider actual cellular factors, not yet known but the study of which should be based on biology and not on psychology. Thus, if a month after a random association, a man presents with a chancre, should the physician consider that he has converted his guilt feelings into an ulceration, or else, as the classical teaching has it, that a treponema, which is not concerned with psychology, has

migrated from one mucosa to another and that this chancre is nothing but a most materialistic bridge-head.

However when Balint states that the onset of a physical ailment creates a new vital situation, and that the adaptation thereto widely exceeds the physiological defence mechanisms, he appears to be quite right. But can one speak of a narcissistic wound, when narcissism itself is only, in the long run, the putting into shape of a deeper and more elemental force? Likewise there is the fundamental notion of a refuge in sickness for those in whom the 'basic defect' of Balint is too intense or extensive and this affords them an unexpected solution. It is true to say that in certain cases 'action' psychiatry attempts forcibly to remove a symptom which only gratifies the patient. However, once again it seems debatable to consider in this way what the author labels collectively as shock treatments, including lobotomy. Surely there could be no question of comparing electric convulsion therapy with lobotomy; and, on the other hand, those who have personal experience of convulsion therapy must admit that in fortunate cases it results in the suppression of a symptom intolerable for the patient himself. This once again is only by the way, but it has seemed of value in so far as the work is almost completely devoid of prejudice. There follows a development which brings clearly worded and precise data regarding the advantages which the patient may hope to receive from the illness, some of which may yield immediate gratification, others a secondary one (such as the importance which the illness gives the subject). But, one may well ask whether these fascinating theoretical considerations have not been developed at the cost of other aspects, usually considered essential, of the psychology of ailing man, namely anxiety and pain. Balint mentions the fear of death, but rather casually, he never

dwells on the repercussions of popular medicine, which surrounds certain disease with a sinister aura. He makes no mention, in particular, of those diseases which, rightly or wrongly, are thought to be social scourges, either because of their transmissibility (as tuberculosis) or because of the conditions of acquiring them, and of the possible consequences thereof (as they were before the modern therapeutic era) as was the case with syphilis. It would appear, on the whole, that it is this chapter, devoted to the relationship of the patient with his disease, which is in greatest need of revision.

One of the best chapters in the book is undoubtedly that dealing with psychotherapy by the practitioner. First of all, it is remarkably forthright, as it recognises the existence of two dangers, the one of considering the physical side of the illness only, and the other, on the contrary, to disregard the physical aspect altogether in favour of the speculative psychogenic roots of the complaint. Above all, Balint teaches great modesty and discretion in the psychological investigations. The practitioner is above all a family doctor and not an amateur psychiatrist; when unsure of himself (which may happen to the most highly qualified) he must be able to listen, and not rush in. By use of clear and striking examples, Balint shows how an enduring assistance can be given to the patient without however violating the sanctuary of his inner feelings.

Lastly in an appendix he deals with another important subject, that of the training of doctors. He feels that practitioners of some standing are more likely to benefit from this kind of teaching than are medical students; they understand the need for it more fully, their experience of life in general is more complete, more personal and especially, the group they belong to is less artificial than is a student's class. Young hospital medical men, how-

ever, of registrar level, may derive benefit from such training, especially if they are working in a general hospital of some importance. This education can not be imparted in formal lectures as, not only is there new information to be conveyed, but it may also be necessary, to some extent, to alter the personal attitude of the student. The ideal system, for Balint, would incorporate a teaching psychoanalysis, followed by formal lectures and by practical work (psychotherapy under supervision). It is obvious that such a method could not be applied to all practitioners; this is why Balint finally restricted his efforts to the third part of the plan, that is psychotherapy under his own supervision and in which it is essentially the counter-transference (that is to say, the feelings of the doctor towards his patient) which has been analysed. Another interesting fact that Balint has stressed is that psychotherapy should be carried out in the very midst of daily general practice, instead of being administered to carefully selected patients. Balint thus shows how, in certain members of his seminar, crises have occurred revealing changes which were taking place in their personality. Above all he insists on the fact, already underlined, that a parallel may be drawn between the relationship of the practitioner and the patient, on the one hand, and that between the group leader and the practitioner, on the other hand. And his investigation, very much to the point, has been concerned mainly with these two aspects, as well as the relationship between

any one member of the seminar and the remainder of the group.

Throughout this whole review, the good points of the book have been shown. One of these, however, has been particularly appreciated, and that is the constant regard for being practical and efficient. Balint is obviously one of those only too rare psychoanalysts who have deigned to alight from the protective nebulae of the freudian Mount Olympus to mix with the common herd. It can indeed be observed that never is a patient faced with the idea of contra-indication to psychotherapy which is certainly in psychoanalysis one of the most likely causes of criticism. By that is meant that, whether psychotherapists or not, psychiatrists should remain physicians whose object is not to effect a cure but to treat according to the rules of the art. The disciples of Balint deal with all comers, and it is likely that they give something to most of them, not necessarily something permanent, but something workable. And again his views on the training of psychotherapist practitioners are also thought out in practical terms. I believe however, that these vital problems could well be, at least, sketched out, at some stage of the university curriculum and this of course, in a live and practical spirit. Just as the formal teaching, *ex cathedra*, of theoretical psychology and even of psychoanalysis would not serve our aim, so a new awareness of the doctor-patient relationship, and the meaning of the illness for the latter, should become part of the education of all doctors.

Man's Posture: Electromyographic Studies

By J. JOSEPH

Springfield, Ill.: Charles C. Thomas, 1960, pp. 88, 44s.

Despite every effort to displace from current use the word 'tone', it survives, and for the good reason that we still have a lingering suspicion that not all the

phenomena ascribed to tone can be explained as reflex responses of muscle. Recent investigations have gone some way to confirm this suspicion: in ascribing

posture wholly to reflex activity—or indeed wholly to muscle activity at all—we were wrong. In this book the established evidence about the mode of maintenance of normal posture is marshalled and new evidence, from a careful electromyographic study, is added. It is shown that, in the standing-at-ease position, much of the postural strain on the back, and all of that on the hips and knees, is taken by the ligaments—and the ligaments are shown to be capable of performing this function. Sustained contraction of muscles may, or may not, occur in addition. Only

in the calf muscles is sustained contraction constantly found: it is argued that, in the standing position, both anterior and posterior ligaments of the ankle are lax, so that muscle contraction is necessary to stabilise the ankle. Postural tone is not, after all, a mysterious entity: it is a combination of simple ingredients.

The reader who is interested in neuromuscular abnormalities will find much that is thought-provoking in this well-written, well-produced little book.

WILLIAM DUNHAM

Illustrating Medicine and Surgery

By MARGARET C. McLARTY

Edinburgh and London: Livingstone, 1960, pp. 167, 37s. 6d.

Miss McLarty's book will be of interest to three distinct groups of people. To those considering taking up medical illustrating as a career it gives a very clear and detailed outline of this type of work and the attributes and training needed for it; to those already embarked on that career it provides refresher material and several tips and ideas that all but the most widely experienced will find new and helpful; and to the research worker and scientist (I put it more widely than 'physician and surgeon') here is a book of reference that will stand them in good stead whether they are contemplating illustrating their own work or want to know the possible ways that a trained medical artist could help to

express graphically the points they wish to make.

Miss McLarty restricts herself to the consideration of illustrations for books and periodicals, demonstrations and lantern slides and touches only lightly on photography and not at all on mounting an exhibition.

The book has 178 illustrations showing equipment, working methods, suggestions for presenting ideas graphically and a full range of medical illustrations executed by the various means discussed. It should be consulted by anyone needing illustrations for an article, a book, a lecture or any other means of teaching or propagating knowledge.

AUDREY BESTERMAN

Structure and Function of the Cerebral Cortex

By D. B. TOWERS AND J. P. SHADÉ

Amsterdam: Elsevier Publishing Company, 1960, pp. 448, 85s.

This book contains the Proceedings of the Second International Meeting of Neurobiologists which was held in Amsterdam in September, 1959. Its appearance in 1960, beautifully printed and illustrated,

is a tribute to the editors and the publishers.

The committee which organised the conference expressed the view that it would be fruitful, at the present time, to

attempt a synthesis of anatomical, physiological and biochemical data on the cerebral cortex; so that it would seem appropriate in reviewing the result to enquire to what extent a synthesis has been achieved, and whether there has been effective cross-fertilisation of ideas between the various disciplines represented at the meeting. The organising body evidently decided that the most appropriate way of achieving these objectives was to divide the meeting into three sections, Neuro-anatomy, Neurophysiology and Neuro-chemistry. In this way 'compartmentalisation' of the three disciplines was effectively maintained. This is a pity since some of the papers, good as they may be, and appropriate as they might have been at another kind of meeting, might just as well have been delivered in the Wilderness, for they were received in silence. The organisers decided to cast their net widely, but one wonders whether a more effective synthesis of views might not have been achieved if they had invited individuals from different disciplines to discuss a few selected aspects of cortical organisation.

One or two topics recur frequently in the different symposia, and one of these is the question of the extent of the extra-cellular space within the central nervous system. On the one hand, electron-microscopists incline to the view that the space between cell membranes is unlikely to be much more than 200 Å, which indicates that the volume of extra-cellular space occupies some 5 to 7 per cent of the total cortical volume; on the other hand, Harreveld and Schadé using a different technique put their estimate of extra-cellular volume in the range of 27 to 35 per cent of the total cellular volume. These great discrepancies may be reconciled, in the view of de Robertis and his colleagues, by assuming that the astrocytes are freely permeable to water and electrolytes, and hence comprise part of the inter-neuronal

water-ion compartment. Things may not, however, be so simple as this, for, as Young points out, there may be a fundamental error in our thinking about the relationships between the cell and its surroundings; he points out that the cell membrane turns in to join the endoplasmic reticulum, so that the space lined by these membranes is in communication with the extra-cellular space, and between the two an interchange of water and electrolytes may thus take place.

The view that some of the glial cells may have a 'trophic' function to the neurones is not new, but it is an hypothesis that is forcing itself into the immediate sphere of interest of the neurobiologist; so that the papers that discussed the structure and function of glial cells were highly topical. In one of these papers, Hyden described an elegant technique for separating, by micro-dissection, neuronal and glial elements from the lateral vestibular nucleus of a rabbit which had been subjected to periodic vestibular stimulation. He found that the ribonucleic acid and enzyme content of the glial cells and neurones were different from comparable material extracted from unstimulated control animals, and he suggested that the nerve cell and the glial cells which surrounded it may possess inter-dependent metabolic systems. Whether or not such an interpretation of these results is justified (and it was criticised by McIlwain and by Towers) Hyden raises issues of considerable importance.

The electrical properties of large populations of neurones, and the effect on a single neurone of operating within such a group, are the subjects of several papers. Bremer discusses, with breadth and clarity, the possible mechanisms that underlie the slow brain rhythms that may be recorded from the cerebral cortex, and Jung of Freiberg and Grafstein of Montreal describe work that has been going on in

their laboratories on the effect of patterned visual stimuli on the activity of single neurones recorded at the visual cortex. The evidence is unequivocal that briskest firing of neurones occurs at positions on the visual cortex that correspond to the edges of the stimulus, and that this behaviour is the result of interaction between many nerve cells. In these experiments very fine electrodes were thrust into the cerebral cortex to record discharge of single cells, but it is not clear from which part of the cell the records were taken. There is abundant evidence that the signal transmitted by an axon is of the all-or-none variety that furnished the basic data of the above experiments, but it is not known whether the cell body and dendrites also show this all-or-none firing characteristic. Various authors have suggested that dendrites do not conduct impulses, and constant reiteration of this belief in recent literature has tended to obscure the tenuous experimental evidence on which this view is based. Eccles, however, in a short but succinct paper reviews the literature on this subject and comes out strongly in favour of the view that dendrites do indeed conduct impulses, though these may have different characteristics from those conducted by the axon.

If the events that go on at the dendrites affect the excitability of the neurone, then the extent and nature of their connections with adjacent nerve cell processes are important in order to understand how signals may be distributed within the central nervous system. Van der Loos gives an account of the dendro-dendritic junctions that are found in the cerebral cortex of the rabbit, and makes some dubious estimate of the number of these junctions present on the basal dendrites of a pyramidal cell. Whatever the number, it would be interesting to know whether this structural proximity implies any functional relationship. Eccles thinks it

does not, for he says, in effect, that if close contact is all that is necessary for functional relationships, there would be chaos within the central nervous system (nevertheless it would still be nice to know whether there is any interaction at such junctions).

It is normally assumed that interaction between adjacent cells operates through the liberation of a chemical transmitter substance at the pre-synaptic terminals. Acetylcholine is one of these substances and Gerebtzoff's paper on the distribution of acetylcholinesterase in Ammon's horn and the cingulate gyrus of rats is of some interest. But one would like to know what the transmitter substance is in the areas in which acetylcholinesterase is absent; and one would like to know the nature of the inhibitory transmitter, not only for theoretical reasons, but for some very practical reasons, such as the role of this substance in the aetiology of epilepsy. With this regard, it is of some interest that Towers finds that some patients with clinical seizures respond dramatically to the administration of gamma-aminobutyric acid, a substance with inhibitor-like properties which is present in certain brain extracts. Thus the inclusion of a paper by Roberts, Baxter and Eidelberg on the metabolism and physiology of this substance is appropriate and valuable.

If the density of neuronal interconnections is of some functional significance at the cellular level, the contribution of Eayrs shows how significant it may be at the macroscopic level of cortical activity (EEG) and behaviour. This paper is a particularly good example of an analysis of the effects of a particular experimental procedure (neo-natal thyroid deprivation) at several biological levels, anatomical, electrophysiological and behavioural and it is to be regretted that so few papers had a similar orientation. Indeed, in only six of the forty-seven papers was behaviour

discussed, a situation that might find its parallel in a symposium on the structure and function of the kidney in which scant attention is paid to the way in which the kidney interacts with its vascular environment.

The discussions that take place in private at a conference, are often more important in terms of clarification and exchange of ideas, techniques and results, than the material that is ultimately published. A reviewer, however, must take as his point of departure the material that appears in print, and, using as evidence the 'Discussion' sections that appear at the end of many of the papers, the extent of such interchanges between individuals from different disciplines was quite limited. On the other hand there was a laudable attempt by Young to achieve some sort of synthesis out of several papers presented in the three symposia. Perhaps a more complete synthesis was not obtained because of the way in which the meeting was arranged; or because (even if these arrangements were perfect) there is just not enough information about cortical

organisation at present available. But it was right to hold the meeting at this time because, as Moruzzi stated in his concluding remarks at the Neurophysiology symposium, there still is, in neurobiology, a common language. Even if this statement is true, it refers to an era that is almost at an end. Soon no-one will be competent to formulate a synthesis because any real communication between the biochemist, anatomist, physiologist, psychologist and clinician will be virtually impossible. Perhaps there ought to be a symposium to decide what to do about this prospect.

Many of the contributions to this symposium reached a high standard and, for anyone interested in the cerebral cortex, this book is well worth reading. To do so however requires a fair working knowledge of anatomy, physiology and biochemistry, and some linguistic competence, since five of the papers are in German and two in French. One wonders what this common language that neurobiologists speak really is.

GABRIEL HORN

Speech Therapy in Cerebral Palsy

By MERLIN J. MECHAM, MARTIN J. BERKO AND FRANCIS GIDEN BERKO

Springfield, Ill.: Charles C. Thomas, 1960, pp. 308, 80s.

This book covers the entire field of the brain injured child and presents a broad survey of much of the information available today on this subject. The book falls into three unequal parts, presumably as contributed by the three authors. This leads to some repetition.

Chapter I deals with the incidence of the condition, its etiology and the description, and classification of the varying symptoms in five groups, spasticity, athetosis, ataxia, rigidity and tremor, with brief mention of developmental problems such as sensory development, perceptual

disturbances, emotional adjustment, intellectual and 'adaptive' development. The neurological development of communication through speech is superficially described.

The second chapter presents a useful approach to the intelligence testing of the brain injured child, in particular stressing the improvement in the I.Q. level of such children with increasing experience and reaction to their environment. The author believes that these children fail specific tests due to specific factors in the brain injury syndrome such, for example, as

perceptual and motor difficulties. He suggests that there is evidence that analysis of Stanford Binet 'scatter distribution patterns' may provide a key for the prediction of the mental development of children with brain injuries. He does not accept the concept that mental retardation is necessarily a fundamental deterrent to speech and language development. Whilst agreeing that language and speech problems may exist when intelligence is normal it must be accepted that the child with limited mental development usually has little thought to communicate to others.

The incidence of disorders of speech in children with brain injury is reported as ranging from 70 per cent to 94 per cent. This is a high figure but probably includes those who are also mentally retarded. Figures are quoted concerning the age of onset of speech. The information in this section suggests that 'cerebral palsy speech' is an isolated phenomenon, whereas it may best be understood in relation to speech disorders in general and as forms of dysarthria involving movement and co-ordination of muscle groups, and disorders of language or aphasia. The speech in these children with brain injury must always show variability dependent on the extent of the language involvement, the motor pathways affected and the child's method of compensation.

The chapter on Speech and Hearing Therapy suggests a 'frame of reference in which the therapist or teacher may utilise his personal resourcefulness.' Various methods of treatment are discussed including relaxation, reflex inhibition, resistive therapy, improvement of neuromuscular control, motor kinaesthetic method, group and individual therapy.

The second section of the book deals with problems of communication in academic learning and may be more useful to the teacher of the child with cerebral palsy as it describes problems concerned

with reading and education. The approach to the child is discussed including his limited experience and interpretation of experience. Problems involved in teaching the child to read and write are described mainly from the psychological rather than the physiological aspect. No mention is made of the use of typewriters for those children whose motor disability makes adequate control of a pencil or pen impossible.

The third section describes language and speech problems from the neurophysiological aspect. The term 'congenital aphasia' is perhaps not very appropriate if it means absence of language from birth. Suggestions for training perception, attentiveness and so forth are given. One is left with the impression that this is an approach to the child through a knowledge of neurology and psychology rather than through knowledge gained by careful observation of the conditions and needs of children. Suggestions are given for the 'sensory integration of language' and 'motor integration for language expression'.

The final chapter describes communication of cerebral palsied children who are severely mentally retarded and discusses intellectual capacity in relation to reading, oral communication and therapy.

Appendix A gives a nucleus vocabulary for Speech Therapy which is somewhat artificial and does not include such first words as 'daddy' or 'mummy'. Appendix B gives the stages of normal verbal language development which may be useful as a yardstick although the wide range of language development in the normal child may suggest that such information should be viewed with discretion.

Appendix C contains well drawn pictures for articulation of sounds in the initial, medial and final position in words, use of which will give some information concerning the child's ability to articulate certain sounds in single words. In children

with cerebral palsy, however, there may be considerable variation in the actual use of such sounds relating to varying bodily postures and emotional circumstances.

This is an interesting book covering a relatively new and wide subject and in

particular speech and language in the child with cerebral palsy. In this area little is yet known with certainty but it contains many suggestions which the discriminating speech therapist can usefully explore.

MURIEL E. MORLEY

The Pathology of Cerebral Palsy*

By ABRAHAM TOWBIN

Springfield, Ill.: Charles C. Thomas, 1960, pp. 206, 64s.

Any medical specialty unsuccessful in arranging a decent marriage with pathology is likely to lead an apologetic sort of life, clinging uneasily to the fringes of scientific respectability. Osler, essaying the study of cerebral palsy in one of his earlier contributions seventy years ago, found its pathology unsatisfactory, 'an outline for our ignorance'. While not specified by Sir William, the failings of the pathology of his day were, charitably estimated, three: barren clinical data, lack of pre- and post-mortem correlations, and total misconception of pathogenesis. Considering the source of material the first of these deficits was inevitable. Specimens came largely if not entirely from institutions providing custodial care for children with major degrees of mental and physical devastation. (Of diplegics Osler remarked: 'The mental condition is profoundly disturbed; the patients are usually imbeciles or idiots, helpless in mind and body'.) Even today this remains true to regrettable degree. The second failing we may venture to hope is in process of correction. But the unquestioned stride forward—taken during the past two or three decades—has been in the matter of pathogenesis. The basic role that anoxia often plays in producing the characteristic lesions of cerebral palsy has been delineated in large measure, in the United States at least, by Cyril Courville of Los Angeles. Anoxia's effects are now further documented in Doctor Towbin's

monograph, his longest and strongest chapter being devoted to the topic.

Having conducted his research partly in central Europe the author uses the local term for the lesions of cerebral anoxia: *elective parenchymal necrosis*. The simplicity of Courville's *anoxic lesions of the brain* may commend itself to American readers. According to Towbin, the hallmark of anoxic brain injury is ischemic neuronal necrosis, which he describes in detail. In shaping his text the author seems to have had in mind the student rather than fellow pathologists. His style is pleasantly clear and unencumbered, and he is sometimes considerate enough to show us the normal structure alongside the illustration of the abnormal. These admirable efforts on the author's part have been abetted by the publisher in providing a well printed, easily handled volume.

In words and abundant photographs Doctor Towbin portrays the late lesions which evolve from ischemic necrosis and the accompanying involvement of glia and blood vessels. From these basic reactions of parenchymal, supportive and vascular tissues are derived the ultimate brain stigmata of cerebral palsy: *convolutional and lobar sclerosis* of the cerebrum, *cerebellar atrophy*, *status marmoratus* of the basal ganglia, and *porencephaly*. Here the writer is at his best, demonstrating the

* An identical review has appeared in *Connecticut Medicine*.

effects of anoxia in a manner to leave no doubt in the reader's mind regarding the part they play in cerebral palsy. The same may be said of the discussions of kernicterus and prematurity.

So far as ensuing material is concerned one would quarrel less with the subject matter than with its ordering. Neuronal storage diseases, the demyelinating diseases and neoplasms do not constitute cerebral palsy according to most definitions, including Doctor Towbin's own ('non-progressive quality . . . is one of the basic criteria'). The author's explanations of these inclusions suggest that he may not be completely happy with the arrangement. Would it not be preferable to gather material of this sort into a separate chapter clearly labelled: *The differential diagnosis of cerebral palsy?*

Doctor Towbin's hand grasps the tiller somewhat uncertainly as he guides us over the treacherous shoals of endogenous and exogenous foetal defect. Cases subsumed under the heading of developmental defect—again a term 'outlining our ignorance'—provide the student with as scant satisfaction as those reviewed by Osler, and for rather similar reasons. This is not the author's fault; such is the state of our science. But like many a lesser mariner Towbin has found irresistible the siren call of irradiation and maternal rubella in pursuing his quest for the sources of cerebral palsy. Of rubella the author says (page 152): 'In children with antecedent intrauterine exposure, mental retardation, motor defects and other central nervous system disturbances are reported'. So far as motor defects of the extremities are concerned this is not true. At least it is not true in the sense implied—that motor sequelae occur in ponderable measure, as is the case with mental retardation and other central nervous system disturbances. In the hundreds of cases of maternal rubella now documented in the literature

foetal motor defects—other than extra-ocular—occur only rarely.

As for irradiation, studies from Nagasaki and Hiroshima have thus far failed to reveal any significant incidence of motor defects (again excepting extraocular) as a consequence of intrauterine exposure of the foetus to atomic bombing. Motor sequelae other than strabismus and nystagmus are in fact so unusual in the children exposed *in utero* to the bombs that one might reasonably conclude foetal irradiation to be an unlikely forerunner of cerebral palsy. While Towbin is unfortunately correct in stating that 'many cases of cerebral palsy have been directly attributed to maternal assault' by rubella and irradiation the reader deserves to be warned that the attributions represent attempts to assimilate to cerebral palsy pathogenic mechanisms known to be concerned in other types of congenital defect. We are dealing here with plausible analogy, not with case reporting. Without question the intrauterine environment can be affected, by man as well as nature, in ways to produce catastrophic effects on the developing foetus. It has yet to be shown that maternal rubella and irradiation ordinarily act in this way to produce cerebral palsy. Indeed, the weight of clinical evidence bearing on the point, which by now is considerable, strongly suggests they do not.

The untutored reader is likely to be led more than a little astray by the heading of the book's last chapter: *The Oligophrenic Triad: Mental Deficiency, Epilepsy and Cerebral Palsy*. Even now cerebral palsy, viewed as a fragment of language, struggles with an overload of semantic ambiguity and misinterpretation. This last unkindest cut is more than it deserves to bear. The relationship among these three diseases, however suggestive and valuable to the synthesizing pathologist, is far from inevitable; each may occur independently

of the others and frequently does. Yet if a case must be made for the linkage there is surely none for equating cerebral palsy with oligophrenia, i.e., mental deficiency.

What the clinician would dearly love to see on his bookshelf is a pathology text to illustrate the status and behaviour of the patients he deals with every day. These children come and go in dispensary and rehabilitation centre, exchange tit for tat with their siblings, take off each day on their special bus to school. The abler among them, perhaps helped by braces or crutches, manage to reach school on their own. They are not so very different from other children—except for arms and legs, that is, and maybe a quirk or two. In adolescence and maturity they arrive at their own compromise with life, finding means of adjustment sometimes startling to their elders . . . Here is the pathology

we should like to know. It is not easy to come by, the literature providing but few examples^{1, 2}. To assemble and annotate material in this sphere the pathologist will require as much help as he can get from clinician and experimental primate laboratory. But Doctor Towbin, possessing great gifts as teacher and writer, is obviously superb pathologist and devoted scientist as well. We may thus expect to find, when a paragon of texts is finally brought to press, that he has been the author, the present volume its first form.

RUSSELL V. FULDNER

¹ Crothers, Bronson and Cobb, S. 'Report of a case of progressive athetosis with lesions in the basal ganglia.' *New Engl. J. Med.*, 1930, 203, 213.

² Gessell, A. and Zimmerman, H. M. 'Correlations of behaviour and neuropathology in a case of cerebral palsy from birth injury.' *Amer. J. Psychol.*, 1937, 94, 505.

Behavioral Change in the Clinic—A Systematic Approach

By GERALD R. PASCAL

London and New York: Grune and Stratton, 1959, pp. 124, 34s.

This is an appalling book, frivolously written, pompous, and pretentious. Its argument is confused and its claims are ridiculous.

Shabby tricks are used to impress the reader with the author's scholarship. The references are inflated to make us think that the author has carefully read the relevant literature and now uniquely combines the thoughts of others in a new and better theory. For example, he writes: 'The student will note as we proceed that our scheme is a hodge podge of Freud, Jung, Horney, Murray, Maslow, Lewin, Tolman, Hull, Hebb, and many others.' I could not see any connection between the hodgepodge set down by the author

and the serious work he listed. I did note that this chapter has 41 references mostly of the kind described.

Another shoddy device is the use of formulae to impress us with the 'scientific' approach. The formulae are written down as though they were magic incantations whose very utterance conferred authority and power on the user. On page 28 we find:

$$PD = f \left[\frac{(S \cdot H \cdot I) \cdot E}{S_r \cdot H_r \cdot P} \right]$$

'in which PD = psychological deficit, S = stress postulated for the particular individual, S_r = stress more prepotent in hierarchy of stresses than S, . . .'

The author then adds: 'The student is

warned that, although the formulation above *seems simple*, the exposition to follow is complex and not always clear, *not even to the author.*' (Italics mine.) Why was this published if it is not clear even to the author?

The main purpose of the book is to set forth a method of psychotherapy based upon stimulus-response psychology. The claim is made that the procedures described are derived from laboratory work. Yet, despite his open antagonism to, and unfamiliarity with, psychoanalysis, the author's third kind of psychological treatment (Type III) has many similarities to psychoanalysis; e.g., he puts his 'subjects' on a couch, the experimenter sits out of view of the subject, the subject may be interviewed as much as 500 times, etc.

Were the author not the director of a psychological clinic in an American university, one could dismiss this book lightly as an effort of no consequence. We are told, however, that the methods described are used on patients in the clinic, and are taught to psychology students in the university. Something ought to be done to protect the students, and, more important, the patients. People who suffer from emotional disturbances and go to a clinic for help deserve to be treated as patients and not as subjects for an experiment. And whatever research value such patients may have is incidental to the main task of a clinic: to provide a clinical service based on humane considerations.

BERNARD ROSENBLATT

Progress in Neurology and Psychiatry

An Annual Review. Volume 14.

Edited by PROF. E. A. SPIEGEL

New York and London: Grune and Stratton, 1959, pp. 656, £4 4s.

It is significant that about a fifth of the pages in this review making up the first of its four parts, deal with recent progress in the basic sciences, including neuroanatomy, neurophysiology and neuropathology. The multitude of new drugs provide material for a chapter on neuropharmacology, with 229 articles reviewed.

The second part of the book covers the progress in neurology, starting with a chapter on clinical neurology, but the work on clinical neurology published in 1958 did not disclose any dramatic new therapeutic advances. Of interest, however, has been the use of corticosteroids as adjuvants in the treatment of bacterial infections of the nervous system.

Among the ten chapters in this second part is one on *Pediatric Neurology* by

Henry W. Baird III, of Philadelphia. He divides the neurological disorders of infants and children into prenatal, perinatal and postnatal groups. In the prenatal group the articles reviewed include some on the infants of diabetic mothers, in whom congenital malformations, including neurological defects, are about twice as common as in the infants of nondiabetic mothers; on phenylketonuria; on cerebral angiomas; and on intrauterine infections, which have become an important cause of meningitis and encephalitis in the newborn. In the perinatal group the new work on jaundice, asphyxia, neonatal meningitis and intraspinal tumours is reviewed. In the postnatal group the articles reviewed deal with a familial syndrome of progressive cerebellar ataxia

oculocutaneous telangiectasia and frequent pulmonary infections, an unusual form of cerebellar ataxia with sex-linked inheritance, pseudotumor cerebri, pseudohypoparathyroidism, polyneuritis and multiple sclerosis in children, and hypsarrhythmia. A syndrome consisting of mental deficiency, bilateral spasticity and ichthyosis, for which a single recessive gene mutation is assumed to be responsible, has already been described in Sweden, but now a case has been reported in a child with German and Italian parents. Epilepsy, the autonomic nervous system, electroencephalography, and the cerebrospinal fluid are other subjects reviewed in this part.

The third part covers the recent progress

in neurosurgery, and the fourth does the same for psychiatry. In this fourth part Sara Dubo and Ralph Rabinovitch survey the publications on child psychiatry which appeared in 1958.

In the 36 chapters of this volume, the 60 contributors give us about 4,000 references from the world's journals and books—a fine bit of work which makes this a valuable and handy reference book for the hospital library. It is therefore remarkable to find not a single article on cerebral palsy. This surely cannot mean that no papers on cerebral palsy worthy of notice were published in 1958?

BO BILLE

Early Diagnosis

Edited by HENRY MILLER

Edinburgh and London: Livingstone, 1959, pp. 400, 25s.

This excellent book has been written for general practitioners by a team of specialists (two in general practice) assembled under the editorship of Dr. Miller of Newcastle. A brisk and instructive introductory chapter by Lord Cohen of Birkenhead sets a spanking pace which is maintained throughout most of the 25 chapters. The use of case-histories and a helpful format make reading a pleasure.

Emphasis throughout is on careful history-taking and meticulous physical examination. Helpful advice is offered on early symptoms and signs and how to detect them. The importance of rectal examination and urinalysis, so often neglected, is given fresh authority. The references to simple laboratory and radiological investigations will be helpful to practitioners who act as clinical assistants in hospitals, and only rarely have the contributors forgotten who they were writing for and wandered upstage among the L.E. cells.

In addition to refreshing memories, this

book adds much of interest. When the history and physical signs are conflicting, the physician (p. 2) discards the history, while the surgeon (p. 18) discards the signs. How is Raynaud to refute the allegation (p. 88) that of the thirty-one patients he described, only one had Raynaud's disease?

The only major sin the book commits is one of omission. Coming from the city of the late Sir James Spence to men who spend so much time with sick children, it is passing strange that there is no paediatric chapter. True, Professor Illingworth has contributed a lucid section on the early diagnosis of Mental Deficiency. But, although nine pages are devoted to disseminated sclerosis, not a word is to be found on haemolytic diseases of the newborn, oesophageal atresia, pyloric stenosis, congenital dislocation of the hip or tuberculous meningitis. It is to be hoped that these and other childhood disorders, the early diagnosis of which may mean so much, will find their way into the next edition.

R. W. SMITHELLS

Essentials of Fluid Balance

By D. A. K. BLACK

Oxford: Blackwell, 1960, 2nd ed., pp. 135, 20s.

Because fluid is as important to life as food, the understanding of electrolyte disturbance and fluid balance is essential to the practice of modern medicine.

Professor Black's second edition is a clear and concise account which leads the reader from Na = Sodium to the intricacies of osmoreceptors, and gives a practical account of the clinical aspects and treat-

ment of the disturbances of fluid balance.

This small book, which is easily read, contains many helpful tables and explanatory diagrams as well as the essential reviews and references required for more detailed study. It can be strongly recommended for those who wish to refresh their knowledge of this subject.

O. D. FISHER

Tools of Biological Research

Edited by HEDLEY J. B. ATKINS

Oxford: Blackwell, 1960, pp. 183, 37s. 6d.

In October 1958 a Symposium was held at Guy's Hospital in which various experts took part with the idea of acquainting surgeons and physicians with many new and unfamiliar scientific tools which may aid them in various research projects. This Symposium has now been published in book form, with an introduction by Sir Cyril Hinshelwood, President of the Royal Society.

The subjects dealt with, by outstanding experts in their own fields, include the design of experiments, followed by flame photometry, electromanometry, tissue culture, tissue transplantation, electron microscopy, weighing cells with the microscope (with some aspects of phase contrast and interference microscopy), electro-

phoresis, image intensification and mass spectrometry. The various experts give a brief historical outline and explain the use of their particular branch of science in a way that should be understandable to readers not fully acquainted with the subjects, but the book may still make slightly difficult reading for those who cannot have had an extensive education in physics and chemistry and other basic sciences. Nevertheless, it would be worth the while of every research worker in any field of medicine to read this book several times, since it opens out new vistas and will help them to plan their researches on a wider base, with better techniques and better tools.

JOHN LORBER

Medical Care of the Adolescent

By J. ROSWELL GALLAGHER

New York: Appieton-Century-Crofts, 1960, pp. 369, \$10.

With the development of Paediatrics as a speciality came the concept of the medical care of human beings by age-group rather than by the particular part of the body which was affected by a disease process. As a result it has been possible in Paediatrics to extend the range of interest

to preventative Paediatrics, then to the newborn, then to surgical Paediatrics, then to normal development (both physical and mental), and now to positive mental, as well as physical, health.

Initially the age up to which the Paediatrician was considered to be legiti-

mately engaged in the study of children was 12 years as an inpatient and 13 years as an outpatient, and this is still roughly accepted in most parts of this country. The reason, of course, is that until that age it is possible to mix the sexes freely, and children's hospitals and units are designed on that principle.

Following Paediatrics came Geriatrics, which also has a close association with social problems and also requires close liaison between hospital and home. As an aspect of general medicine Geriatrics seems to have gained a place in the medical community which is likely to persist. Nothing, however, has come so far of Sir Heneage Ogilvie's suggestion that the study of young Service men and women should be called 'Ephebiatrics' with, presumably, its unpronounceable practitioner, the Ephebiatrician! Nevertheless, in Boston over recent years a unit has been formed which would fulfil some of the requirements of such a specialty since it studies the age-group of the adolescent which is defined in Dr. Gallagher's book as being from 12 to 21 years inclusive. It is, therefore, the study of that specific age-group which follows the one normally seen by the Paediatrician, and the flavour of this book is that of Paediatrics.

Apart from the disorders of puberty there are, of course, very few physical abnormalities which occur only at adolescence, and in this way the period 12-21 years differs from the 0-12 years of Paediatrics, but some conditions certainly tend to start at this time (e.g., diabetes) and many tend to improve. On the physical side, therefore, there is not so much to be learnt from the study of this age-group, although Dr. Gallagher makes the important point that severe physical exercise

starts at this age and sometimes requires medical (and surgical) advice.

In the excellent series of essays which compose the book, Dr. Gallagher clearly views himself and his associates as the overall doctor for the adolescent and the one who sees the child of that age-group as a whole person. He has with him a large number of experts in a wide variety of regional specialties, and his book reflects the concept of the unity of the adolescent whatever disease process afflicts him. The subjects covered will interest physicians, orthopaedic surgeons, gynaecologists, psychiatrists and paediatricians, as well as school medical officers and even student health officers. In all the chapters the author stresses the importance of viewing the adolescent as a special being, neither child nor adult. This is assuredly correct and there seems no doubt that more of this type of thinking is required in this country as well.

To pick out any particular essay as outstanding would not be particularly helpful but one alone reflects, perhaps, the author's deep understanding of the tempestuousness of the adolescent period, and this chapter's title, *Sex, Homesickness, Rebellion, Anxiety and Delinquency*, seems to sum it up most aptly.

The Adolescent Unit at Boston, of which Dr. Gallagher is the chief, is renowned throughout the world as a bold enterprise. It is likely to be followed by others for whom this book will become basic equipment. For all others who have to deal with adolescents it can be thoroughly recommended. Unfortunately there is no essay on the adolescent with cerebral palsy, but this oversight can easily be remedied in future editions.

R. R. GORDON

ABSTRACTS

In collaboration with *Abstracts of World Medicine*, published by the British Medical Association, and with the kind assistance of the Excerpta Medica Foundation *Courier*, and *Obsterical and Gynecological Survey*.

The Natural History of Infantile Spasms

P. M. JEAVONS and B. D. BOWER. *Archives of Disease in Childhood*, Feb., 1961, **36**, 17-22, 14 refs.

As a preliminary to a study of the effect of corticotrophin in children with infantile spasms and mental retardation, for which good results have been reported, the authors, working at the Children's Hospitals and University of Birmingham, undertook this survey of the progress of 30 such children who had received no hormone treatment. All had been treated with the common anticonvulsant drugs with little effect and 14 had had a course of one of the tetracycline drugs, with unconvincing results, and the authors consider that for the purposes of this study the patients could be regarded as untreated.

In all cases the spasms had appeared before the age of 12 months; in 15 (the symptomatic group) some aetiological factor was found, in a further 2 the condition was of doubtful origin, but in the others it was cryptogenic. All had undergone clinical and EEG examinations at about 6-monthly intervals for 2 to 6 years. By the age of 12 months 3 were free of spasms, at 36 months 16 were free, and at 42 months only one-third of the 26 survivors still had spasms; the frequency and severity of the spasm also decreased with advancing age. The symptomatic and cryptogenic groups showed little difference, except that 6 of the former showed later-

ling signs while none of the latter did so. The EEG, recorded in 25 of the patients under the age of one year, was abnormal in all, being highly disorganised in 13, less so in 4, while 8 showed epileptic characteristics; however, at age 30 to 36 months the EEG record had become normal in 10, was still epileptic in 8, but was now highly disorganised in only 3. The tracing also improved with age and there was a close correlation between clinical condition and EEG findings. The cryptogenic group showed fewer disorganised EEG records throughout the period than the symptomatic group, and the abnormal recordings returned to normal earlier.

No general mental improvement was found; at 12 months 16 of the children were grossly retarded and 13 moderately so, while at 42 months 11 were still grossly retarded, 12 moderately retarded, and one normal. In this last child and in another who became normal at 36 months the initial EEG had shown few abnormalities and was normal on second and subsequent recordings, while the spasms ceased at 7 and 20 months respectively; one of these 2 patients had received tetracycline. At 12 months and subsequently the symptomatic group contained a higher proportion of grossly retarded children, whereas patients in the cryptogenic group showed a slight tendency to improve, so that at 42 months none were grossly retarded and 2 were normal mentally.

Janet Q. Ballantine

The Effect of Corticotrophin and Prednisolone on Infantile Spasms with Mental Retardation

B. D. BOWER and P. M. JEAUVONS. *Archives of Disease in Childhood*, Feb., 1961, **36**, 23-33, 19 refs.

In this further study the authors have investigated the effect of treatment with ACTH or prednisolone in 23 children with infantile spasms and mental retardation, of whom 11 were classified as symptomatic and 12 as cryptogenic (as previously defined). The EEG initially abnormal in all, was recorded twice weekly while the patients were in hospital (4 to 8 weeks) and as often as convenient during the follow-up period. The mental status was assessed in all cases before treatment, 3 weeks after its completion, and at intervals during follow-up. Treatment was with either 20 to 30 i.u. of ACTH-gel daily, given every 12 hours, or prednisolone in a dosage of 1 mg. per lb. (2.2 mg. per kg.) body-weight daily in 4 divided doses orally. The maximum dose was continued as long as the EEG tracing was improved, and thereafter the dosage was gradually reduced. A second course was required in 8 cases for relapse or lack of response to the first, and one child received a third course.

In the cryptogenic group of 12 children the spasms were controlled in all by the first course of treatment, in 5 cases in the first week but in 2 not until after 5 weeks; however 8 patients relapsed within 3 weeks of the end of treatment, usually in the first week. One child also had major fits which persisted during treatment and on discontinuing the drug status epilepticus developed. The 8 who relapsed were given a second course, but again relapsed. The EEG showed definite signs of improvement in all patients during the first course, and in 6 the record became non-epileptic, 3 of these showing completely normal records thereafter. No relationship was apparent between cessation of spasms and

the EEG tracings; both usually became normal or nearly so within a week, but appeared to do so in random order. A non-epileptic record was obtained any time between 7 and 64 days, a time unrelated to dosage or type of drug. No immediate improvement in mental development was noted and 4 patients appeared to deteriorate. Of the 5 given a second course and the 1 given a third course only one patient responded to the longer drug treatment, in the form of mental improvement. Follow-up at 3 to 14 months showed that 2 more patients were free of spasms, making 7 in all. Mental testing showed a rise of over 10 per cent in the development quotient (D.Q.) in only 3 cases, the others being unchanged.

In the symptomatic group, steroid treatment produced only temporary relief of spasms and improvement in the EEG, except in one patient in whom the improvement was maintained and the D.Q. had risen by 20 per cent at follow-up. The combined results of the two groups showed that during treatment the spasms ceased in 81 per cent and the EEG improved in 95 per cent, becoming non-epileptic in 39 per cent, but that 15 patients relapsed within 3 weeks of stopping the drug. The later effects of treatment were shown by considering children who had completed treatment more than 3 months previously. Of these, 3 out of 5 aged 12 months were free of spasm (only 3 out of 29 untreated patients were free at this age) and at 18 months 2 out of 6 were free (6 out of 29 in the untreated group).

The authors consider corticotrophin to be slightly more effective than prednisolone in the dosage used in this study. They did not find that early treatment was important, and were unable to confirm the finding of Sorel and others that rapid mental improvement followed treatment with corticotrophin. Only one patient who had a highly disorganised EEG

initially became normal mentally with a normal EEG; otherwise the degree of mental improvement was not significantly greater than in the untreated group.

Janet Q. Ballantine

The Electroencephalogram in Neonatal Convulsions

R. HARRIS and J. P. M. TIZARD. *Journal of Pediatrics*, Oct., 1960, 57, 501-520. 25 figs., 35 refs.

The authors describe the electroencephalographic abnormalities in 41 infants in whom convulsions developed in the neonatal period and then attempt to classify these abnormalities and correlate them with the clinical state. Of the 41 infants, 7 died in the early neonatal period, and 31 of the survivors were followed up for a year or longer. It was not always possible to make an exact diagnosis of the primary condition underlying the convulsive state; nevertheless, they list the clinical states associated with fits in these infants, perinatal anoxia being the most frequent. They point out that it is not always easy to recognise fits in the newborn or to distinguish apnoea causing a fit from a fit causing apnoea. However, most of the fits were either tonic, characterised by symmetrical tonic contractions with opisthotonos, extension of the arms and legs, and sometimes followed by clonic movements; or focal, with localised jerking. The EEG abnormalities included: (1) rhythmic slow waves; (2) persistent focal sharp waves; (3) spikes; (4) repeated stereotyped sharp waves or wave complexes; (5) gross asymmetry; (6) small amplitude; (7) sharp waves during episodic sleep activity; and (8) fast activity.

The EEG was recorded during seizures in 18 infants; in 5 of those with localised clonic fits there were repeated focal stereotyped complexes arising contralaterally, while in 8 with tonic fits there was

flattening only. The duration of the convulsive illness seemed to be the most important factor in assessing prognosis, although inter-seizure EEG abnormality was more often seen in the patients who did least well.

The authors consider that if the practical importance of the EEG in the newborn infant appears slight at present, its theoretical implications are of the greatest value.

N. S. Alcock

Epilepsy and Television

C. MAWDSLEY. *Lancet*, Jan. 28, 1961, i, 190-191, 12 refs.

The author reports on 3 cases of television-induced epilepsy seen at the Manchester Royal Infirmary during 1959. The fact that flickering light can cause epileptic seizures has been known since Roman times. Recently electroencephalographic investigations have augmented clinical observations, and the author briefly reviews the literature to illustrate this point. There were marked EEG changes in all 3 cases reported, and 2 of the patients had fits when stimulated with the stroboscope. The onset of the epileptic attacks is described and details of the EEG findings are given. The flicker frequency of a faulty television set is likely to be 25 per second, a rate which corresponds to the frequency at which atypical spike discharges and fits were elicited experimentally. There was a consistent association between fits and television flicker in these cases. All 3 patients were given anticonvulsants and advised not to watch a flickering screen. (The author does not discuss whether avoidance of photic stimulation alone would prevent the fits, or whether anticonvulsants could suppress the fits even in the face of a flickering screen.)

M. R. Medhurst

Clinical Course of Hyperbilirubinemia in Premature Infants

K. HUGH-JONES, J. SLACK, K. SIMPSON, A. GROSSMAN and D. YI-YUNG HSIA. *New England Journal of Medicine*, Dec. 15, 1960, 263, 1223-1229. 5 figs., 39 refs.

The natural history of hyperbilirubinaemia, which is frequently observed in the premature infant, was studied at Cook County Hospital, Chicago, in 122 such infants weighing 2 kg. or less at birth who were not treated by exchange transfusion. The degree of hyperbilirubinaemia varied with the degree of maturity, the serum bilirubin level being highest in the smallest infants and in those whose gestation period had been the shortest. Kernicterus was diagnosed in 3 infants, the diagnosis being confirmed at necropsy in 2 of them. The serum bilirubin level was over 20 mg. per 100 ml. in these 3 infants, who represented 6.7 per cent of the total infants in the series with a peak bilirubin level exceeding this value and 30.5 per cent of those in whom the level exceeded 30 mg. per 100 ml. Neurological abnormalities were observed during the first week of life in 19 (42.2 per cent) of the infants with a serum bilirubin level over 20 mg. per 100 ml. and in 18 (23.4 per cent) of those in whom the level was below 20 mg. per 100 ml. These signs did not form the classic triad of kernicterus—namely, opisthotonos, shrill cry, and absent Moro reflex—and did not persist as the children grew older.

The authors recommend exchange transfusion in premature infants with a serum bilirubin level of over 20 mg. per 100 ml.

Winston Turner

The Drug Treatment of Icterus Gravis Neonatorum (In German)

G. WOHLMUTH and P. KISS. *Acta paediatrica Academiae Scientiarum Hungaricae*, 1960, 1, 41-54. 4 figs., 35 refs.

In recent years various drugs have been used in addition to, or as a substitute for,

exchange blood transfusion in the treatment of haemolytic disease of the newborn. During the past 2 years the authors, working at the Municipal Hospital for Premature Infants, Budapest, have employed this form of therapy in 206 cases of icterus gravis in premature babies; in only 16 of these cases was exchange transfusion necessary in addition. The two main drugs used were cortisone and polyvinyl-pyrrolidone (polyvidone). The latter acts by combining with bilirubin in the serum, the resulting chemical complex being excreted by the kidneys, while cortisone has an inhibiting action on the antigen-antibody reaction, renders erythrocytes less permeable to antibodies, and has a protective action on the liver and nervous and collagen tissues. The indications for use of drug therapy in these cases are discussed.

The treatment so far seems to have had no undesirable side-effects. Since its introduction there has been a considerable fall in the mortality from kernicterus, but the authors agree that final judgment must be postponed until a long-term neurological follow-up study can be carried out.

Marianna Clark

Glucose-6-phosphate Dehydrogenase Deficiency: a New Aetiological Factor of Severe Neonatal Jaundice

S. A. DOXIADIS, PH. FESSAS, and T. VALAES. *Lancet* Feb. 11, 1961, i, 297-301, 22 refs.

Between January, 1957, and June, 1960, at the Alexandra Maternity Hospital, Athens, 220 babies received exchange transfusions and a further 30 were admitted with established kernicterus; in 105 of the former (46 premature) and 25 of the latter (2 premature) neither Rh nor ABO blood-group incompatibility was present. Of the 82 full-term infants with no incompatibility 40 were first-born, and in the 42 families with a previously live-born infant 11 of the siblings had been affected with severe neonatal jaundice. It therefore

seemed possible that some hereditary haemolytic factor was the cause of the jaundice, and in view of the frequency of favism in Greece, the authors suspected that a deficiency of glucose-6-phosphate dehydrogenase might be responsible. The activity of this enzyme in the erythrocytes was therefore determined by the method of Motulsky and Campbell in: (1) cord blood from 100 newborn infants collected at random; (2) the blood of all infants born in or admitted to the hospital with neonatal jaundice of sufficient severity to warrant serum bilirubin estimation and also that of their parents; (3) the blood of surviving members of families with a history of severe unexplained neonatal jaundice.

Normal enzyme activity was found in all infants in Group 1, but results in the other two groups revealed an appreciable number of infants and families with a genetically transmitted defect of the enzyme. It seems clear that in Greece, and probably also in some other countries, glucose-6-phosphate dehydrogenase deficiency accounts for some cases of severe neonatal jaundice and kernicterus. It is noted that this variety of jaundice is not always severe on the first day of life; its appearance is an indication for exchange blood transfusion.

F. P. Hudson

5-Hydroxyindoles in Mental Deficiency

C. M. B. PARE, M. SANDLER and R. S. STACEY. *Journal of Neurology, Neurosurgery and Psychiatry*, Nov., 1960, **23**, 341-346. 1 fig., 19 refs.

In a previous study (*Arch. Dis. Childh.*, 1959, **34**, 422; *Abstr. Wld Med.*, 1960, **27**, 409) the authors observed that some non-phenylketonuric mental defectives had abnormally high serum levels of 5-hydroxytryptamine (5-HT). In the present, more extended, study they have therefore determined the serum and platelet 5-HT content, platelet adenosine triphosphate

content, and level of urinary 5-hydroxyindoleacetic acid (5-HIAA) and urinary creatinine excretion in 83 non-phenylketonuric mentally subnormal patients drawn from four London hospitals and 68 control subjects, of whom 16 were normal adults and children. The method of determining the capacity of platelets to absorb 5-HT (devised by Stacey but not yet published) consisted in incubating platelet-rich plasma at 37° C. for 90 minutes with excess 5-HT in an atmosphere of 5 per cent carbon dioxide. As no significant differences were found between the normal adults and normal children in regard to 5-HT and 5-HIAA excretion per g. urinary creatinine these two sub-groups were amalgamated.

The findings in the defective groups appeared to be independent of the hospital of origin and could not be correlated with any effect of drugs. In 12 cerebral palsy patients with average intelligence the mean serum 5-HT level was normal, but in 24 out of 28 such patients with an I.Q. of less than 50 it was considerably and significantly raised. Of 10 mongol children 9 showed a normal serum 5-HT level, but in the tenth it was 257 $\mu\text{g.}$ of 5-HT per ml. of serum, the mean control value being $145 \pm 13 \mu\text{g.}$ In 6 cases of tuberous sclerosis (epiloia) it was, at a mean of 215 $\mu\text{g.}$ per ml., around the upper limit of the normal range, according to the text (but it is shown as significantly different ($P < 0.05$) in the authors' table). In 4 patients whose mothers had had rubella during pregnancy the level (391 $\mu\text{g.}$ per ml.) was significantly above the normal. In single cases of the Sturge-Weber syndrome, gargoylism, and probable cerebral lipidosis, the value was also significantly high, but in single cases of hypertelorism and ichthyosis with spastic paraplegia it was normal. In a group of cases of unclassified mental deficiency the serum 5-HT level was above the normal

range in 20 while in 11 it was within it (the authors' table shows a total of only 30 such cases). The results for numerous other small groups, each of 2 or 3 cases, are also reported (but for these the original paper should be consulted). Attempts to establish correlations between the serum 5-HT level and various aetiological factors and also such factors as degree of physical handicap, intelligence, nutritional status, or the presence or absence of epilepsy in mental defectives were not very fruitful.

In the patients the platelet 5-HT concentration after incubation reached the same level as that in the controls, although before incubation this value had been about $2\frac{1}{2}$ times that in the controls. In 4 cases with a high serum 5-HT level the platelet adenosine triphosphate content was no higher than in 5 normal subjects.

(In that part of the paper dealing with urinary 5-HIAA excretion only the ratio of 5-HIAA to creatinine is given. As the authors did not know if creatinine excretion was normal, the daily amount of 5-HIAA excretion is unknown. In addition, the authors state that 'overnight urine samples were used when possible, but many of the patients were incontinent and with these random samples were used . . . no precautions were taken to control dietary intake of 5-hydroxyindoles. For these reasons less reliance can be placed on the 5-HIAA than on the 5-HT figures'. Consequently, the abstracter has ignored these figures.) *G. de M. Rudolf*

Environmental Treatment of a Hereditary Illness: Wilson's Disease

I. H. SCHEINBERG and I. STERNLIEB. *Annals of Internal Medicine* Dec., 1960, 53, 1151-1161, 32 refs.

The inherited defect in patients with hepatolenticular degeneration (Wilson's disease) is the inability to synthesize a normal amount of the plasma copper-

protein ceruloplasmin; this is a blue globulin which is normally present in the plasma in a concentration of approximately 30 mg. per 100 ml. Marked and permanent deficiency of ceruloplasmin, which occurs only in Wilson's disease, results in the deposition of abnormal amounts of dietary copper in the tissues.

This paper from the Albert Einstein College of Medicine, New York, describes an attempt to treat the disease by the intravenous administration of a purified, concentrated preparation of ceruloplasmin made from human plasma. The preparation was first tested on 2 control subjects without untoward results and was then administered intravenously to 2 female patients, aged 24 and 18, who had had manifest Wilson's disease for the previous 5 years, the serum levels of ceruloplasmin being less than 1 mg. per 100 ml. After the infusion the serum ceruloplasmin level rose to about 30 mg. per 100 ml. and remained over 15 mg. per 100 ml. for 4 to 5 days. During this period however, no effect was produced on the faecal or urinary excretion of orally administered radioactive cupric (^{64}Cu) ions, and no ^{64}Cu ions combined with ceruloplasmin. No clinical effects, beneficial or otherwise, were observed.

At present, therefore, treatment must be directed to the 'environment copper'—that is, by reducing the intake of copper and promoting the excretion of copper already deposited in the tissues. To this end the authors give: (1) a diet low in copper content; (2) potassium sulphide, 40 mg. orally, with each meal, since this substance precipitates, and thereby renders unabsorbable, copper in the diet; and (3) penicillamine in a daily dosage of 1 to 4 g. in divided doses, since this amino-acid, by combining with copper in the tissues, increases the urinary excretion of copper. To combat the iron deficiency which may result from this regimen,

ferrous sulphate is given on one day per week, the potassium sulphide and penicillamine being omitted on this day.

The results of this 'de-copperising' treatment in 10 patients for periods ranging from 1 to 6 years are described. Kayser-Fleischer rings, which were prominent in 2 patients, disappeared under the treatment. However, liver function, as estimated by clinical and laboratory tests, improved in only one patient, and one died of liver disease. Neurological improvement was striking in several patients. It is important, therefore, to diagnose potential patients by screening tests before they become symptomatic and to begin treatment while they are still under one year old.

Joseph Parness

Correlation Between Birth Weight and Clinical Findings in Diplegia

E. M. RUSSELL. *Archives of Disease in Childhood*, Dec., 1960, 35, 548-551. 3 figs., 11 refs.

In this study the author has analysed the birth weights and clinical findings in 200 diplegic children aged from 14 months to 13 years, 58.5 per cent of them being males, referred to the Edinburgh Clinic of the Scottish Council for the Care of Spastics. The distribution of birth weights in the general population shows a unimodal curve. Among these children, paraplegics whose upper limbs were functionally normal showed a bimodal curve with a major peak at 4 lb. (1.8 kg.) and a smaller peak at 8 lb. (3.6 kg.). In the case of triplegics and tetraplegics the curve was also bimodal, with a main peak at 7 lb. (3.17 kg.) and a lesser rise at 3 lb. (1.36 kg.). Paraplegics thus tended to be premature while those with upper limb involvement tended towards average weight.

Assessment of the intelligence showed that paraplegics as a group were more intelligent than the triplegics or tetraplegics, the difference being statistically

highly significant ($P = < 0.001$). When the patients were further subdivided into premature and mature, it became evident that those with the smaller birth weights tended to be more intelligent than those with the larger, the difference in the case of paraplegics being significant ($P = 0.01$); however, no significant difference was found between premature and mature triplegics and tetraplegics. The incidence of epilepsy among paraplegics was 9 per cent, compared with 26.8 per cent in the others. Strabismus showed no significant difference in incidence in the two groups. The mean birth weight in patients showing mental impairment and/or epilepsy was 6.1 lb. (2.73 kg.) whereas in those in whom both these were absent the mean was 5.0 lb. (2.27 kg.).

The author discusses the significance of her findings with reference to the possible differing aetiology of the diplegic condition in premature and mature babies.

Janet Q. Ballantine

The Treatment of Muscular Spasticity of Central Nervous Origin with a Benzodioxan Derivative ('Quiloflex')

J. KRISCHEK and K. SUWELACK. *Münchener medizinische Wochenschrift* Dec. 23, 1960, 102, 2597-2598, 10 refs.

This assessment of 'quiloflex', a new relaxant for striated muscle, is reported from the University Neurological Clinic, Münster, where the authors have used the drug in the treatment of 51 patients suffering from a wide variety of central nervous disorders in which increased muscle tone was the main symptom. Most of the patients had been ill for over a year, though the duration of the illness ranged from 2 weeks to 26 years. The drug was assessed over a period of 1½ years. The diagnosis and number of cases in each category with the results graded as good, fair, and negative are tabulated. Medication was started with small doses

and gradually increased until the optimum effect was achieved. Length of treatment and rapidity of response varied greatly; in most cases the therapeutic effect—diminution of muscular spasm—was noticeable within 5 to 7 days, but in some not until after 4 weeks. Side-effects, which occurred in 22 patients, consisted in faintness, giddiness, perspiration, nausea, vomiting, and headache, but the most troublesome was weakness of the legs. In most cases these symptoms subsided when the dosage was temporarily lowered. It is stated that suppositories of the drug are now available which should help to abolish the gastric side-effects.

Quiloflex was found to produce symptomatic relief only, symptoms recurring when it was discontinued. During the trial it was found that the drug enhanced normal remissions and that it facilitated physiotherapy. On the basis of satisfactory results in 38 of their 51 patients the authors suggest that this drug is a useful addition to the treatment available for muscular spasticity.

M. R. Medhurst

The Early Diagnosis of Foetal Asphyxia. (In Russian)

A. M. ARNOLDOVA. *Voprosy ohrany materinstva i detstva*, Jan., 1961, 6, 48–51, 18 refs.

Success in dealing with intra-uterine asphyxia depends on early diagnosis and prompt treatment, but the early signs of this condition are not clearly defined in the current literature. The author has therefore investigated 114 pregnant women, of whom 66 were healthy, 21 had late toxæmia of pregnancy, 14 cardiovascular disease, 9 hypochromic anaemia, and 4 other pathological conditions.

In the healthy women the maximum arterial blood-pressure was 120 and the minimum 60 mm. Hg, the erythrocyte-count 3 to 4 million per c.mm., and the haemoglobin value 50 to 70 per cent.

The normal oxygen saturation of arterial

blood varies from 88 to 96 per cent. Hench's test (breath-holding in expiration) leads to an increase in the maximum arterial blood-pressure of 8 to 10 mm. Hg, an increase in the maternal pulse-rate, and slowing of the foetal heart rate by 4 to 6 beats per minute. During the test the oxygen saturation of the maternal arterial blood fell in primiparae by 10 to 15 per cent and in multiparae by 15 to 16 per cent, returning to normal in 5 to 10 seconds in the former and in 16 to 20 seconds in the latter. Breath-holding lasted an average of 30 seconds in the primiparae, but only 20 seconds in the multiparae.

In 18 women with nephropathy the blood-pressure was between 160 and 180 mm. Hg; oxygen saturation was below 70 per cent in 10 patients, and between 70 and 80 per cent in 5. Hench's test led to a slowing by 8 to 10 beats per minute in the foetal heart rate, and the above 15 patients could hold their breath for only 15 to 20 seconds. The 9 women with hypochromic anaemia gave even more positive evidence of low oxygen saturation, and during Hench's test the foetal heart became arrhythmic in 5 cases. Similar results were obtained in the patients with cardiovascular disorders. Oxygen therapy, especially when administered subcutaneously, improved the oxygen saturation and restored the foetal heart rate to normal; it is considered to be a life-saving measure for both mother and child. In 5 cases in which the foetal heart sounds were inaudible or nearly so and in 2 very serious cases of foetal asphyxia due to maternal nephropathy oxygen therapy permitted the delivery of living children.

The author concludes that Hench's test is a very valuable clinical indication of the mother's adaptability to oxygen insufficiency.

L. Firman-Edwards

A New Autosomal Trisomy Syndrome: Multiple Congenital Anomalies Caused by an Extra Chromosome

D. W. SMITH, K. PATAU, E. THERMAN and S. L. INHORN. *Journal of Pediatrics*, Sept., 1960, 57, 338-345.

The authors report, from the University of Wisconsin, Madison, the cases of 2 babies possessing an extra autosome in the 16 to 18 group. The infants, one male and one female, died of cardiac failure at 2 and 2½ months of age respectively. The clinical features common to both were probable mental defect, spasticity, micrognathia, malformed and low-set ears, umbilical hernia, intraventricular septal defect, and patent ductus arteriosus. The mothers were both aged 46 at the time of conception.

The authors have since seen 4 further children, all unrelated, with the same syndrome, which they now interpret as

being trisomy for chromosome 18. They also note that this syndrome appears to be the same as that described by Edwards *et al.* (*Lancet*, 1960, i, 787). C. O. Carter

Note: Clinically, the syndrome described by Edwards and his colleagues is similar to that described by Smith and his collaborators. The child described by Edwards had a webbed neck and some clinicians would diagnose this as Bonnevie Ullrich's syndrome. In this patient the interpretation of the chromosome analysis was that she was trisomic for chromosome 17. With the present uncertainty of chromosome identification, all one could now say is that children with the syndromes under discussion have been described who are trisomic for one of the chromosomes in the 16-18 group, most likely chromosome 17 or 18 of the Denver nomenclature.—

Paul E. Polani

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Under the eye of the Prince Consort, looking down from a wall in the German Hospital, London, Dr. Parkes Weber is presented with a copy of his *Collected Writings* by Dr. C. K. Blum, who edited them. The occasion was Dr. Parkes Weber's 80th birthday. The other figures in this photograph are Mrs. Parkes Weber and Mr. Hugo Rast, hon. senior surgeon to the hospital.

PERSONALITIES OF TODAY

Frederick Parkes Weber

ON May 8, 1961, Dr. Parkes Weber celebrated his 98th birthday. He became physician to the German Hospital in London in 1894 and to the Mount Vernon Hospital for Chest Diseases in 1899. He had made a reputation for himself in the discussions of half a dozen or more medical societies by the later 'nineties. He was president of the Assurance Medical Society in 1918, first Mitchell Lecturer of the Royal College of Physicians in 1921 and Moxon Gold Medallist (Clinical Research) in 1930. To quote a notice in the *British Medical Journal* on the occasion of his 90th birthday, 'What has distinguished him during all these years has been his zeal for clinical studies, his search for the more enigmatical signs and symptoms of disease, and the clarity with which he had expounded what he has discovered.'

On his 80th birthday, his friends at the German Hospital presented him with a volume of his Collected Writings. It could be the titles only, for the separate writings ran to nearly a thousand, beginning in 1890 with a contribution to *St. Bartholomew's Hospital Reports* (he was then house-surgeon at St. Bartholomew's) on abnormal foramina in the heart and its valves, and ending in 1943 with a short paper in the *British Medical Journal* on congenital jaundice in a man of 77.

Since then, the spate of books and other original contributions to medical journals has not ceased to flow.

Some of his own original case-reports and nosographic observations have led to his name being attached eponymically to several disease states: hereditary haemorrhagic telangiectasia of the skin and mucous membranes;¹ Rendu-Osler-Weber disease; haemangiectatic hypertrophy of the limbs; Weber-Klippel syndrome;² the syndrome of haemangiectatic naevoid conditions involving both face and cerebral meninges; Sturge-Weber-Kalischer disease;³ and relapsing febrile nodular non-suppurative panniculitis, afterwards known as Weber-Christian disease.⁵ Examples of his earliest full clinical reports on commoner conditions include 'Chronic Polycythaemia with Enlarged Spleen, probably a disease of the Bone Marrow';⁶ 'Arteritis Obliterans of the Lower Extremity'⁷ (before Buerger's paper entitled 'Thrombo-Angitis Obliterans';⁸) and 'Auriculo-Temporal Syndrome', or Frey's syndrome.⁹ His very full report on a typical example of Cushing's pituitary syndrome with a basophil adenoma¹⁰ was included in Cushing's original series in 1932¹¹. A list of even some of his more significant medical observations would far outrun the scope of a brief biography.

A fellow nosographer with Osler,

Christian and Cushing, he survives as an inspiring example of the classic scholar physician. His studies of developmental and congenital disorders of the brain (e.g., the Sturge-Weber-Kalischer syndrome) and his pioneer studies of word deafness¹² and anosognosia¹³ entitle him to a special place in this journal.

Dr. Parkes Weber has been a fellow of the London Society of Antiquaries since 1891, a fellow of the Royal Numismatic Society since 1885, and a member of the Cambridge Antiquarian Society since 1883. Many honours in this second field will shortly culminate in 'Parkes Weber Day' at the British Museum on November 18. This other interest provides an interesting clue to Dr. Parkes Weber's contribution to medicine. He is a great collector and even a public apologist for collecting.¹⁴ As a collector, it was natural for him to seek rarities, and this necessitated special interest in nomenclature. He has defended his interest in rare diseases and rare cases by quoting Sir James Paget:¹⁵

'... we ought not to set them aside with idle thoughts or idle words about curiosities or chances. Not one of them is without meaning; not one but might be the beginning of excellent knowledge, if only we could answer the question: Why is this rare? Or, being rare, why did it in this instance happen?'

As a collector, he acquired an encyclopaedic knowledge of the world's medical literature of his time, which, together with his vastly broad and long experience, has often enabled him to resurrect nosographs from forgotten limbo in a way that has been salutary if a

little embarrassing for the 'rediscoverer'. The delight with which he has carefully observed a tiny detail of the often intentionally enigmatic design on a coin or amphora has been well communicated in his papers. He has never been a miserly collector, having given away outstanding collections to the Boston (U.S.A.) Medical Library, the Bodleian at Oxford, the Fitzwilliam Museum in Cambridge, and the London Guildhall Library.

One of his favourite physical recreations has been mountaineering; he has led a good many expeditions and climbed the Breithorn at the age of 70. He used to walk daily through the London *Blitz* from the West End through the back streets of St. Pancras and Islington to the German Hospital in Dalston, and remained physically active until 1948 when he fractured his femur in crossing Cavendish Square.

His father, Sir Hermann Weber (of Weber's neurological syndrome), was acclaimed for all-round contemporary clinical repute and variety of personal experience by Sir Henry Bashford in 1951, in his study of fellows of the Royal College of Physicians.¹⁶ In his 80s his son stimulated his interest in coin-collecting to such effect that Sir Hermann's collection became a valuable asset to the British Museum.

Parkes Weber was born in 1863, educated at Charterhouse and Trinity College, Cambridge, studied clinical medicine at St. Bartholomew's Hospital, London, and married Hedwig Laissle, a promising paediatrician, in 1921. Theirs has been a wonderfully happy marriage, and his wife helps him to

keep in close touch with medical progress despite his physical disabilities. Thanks to her, he can still reply constructively to the numerous enquiries he receives about baffling cases allied to those he has meticulously observed and recorded in the past. With her help, he remains the last international court of appeal about medical rarities and the history of special investigations, such as lumbar puncture, etc. As befits an observer of life of his eminence and experience, he has not neglected philosophy, comparative religion and dialectic in his writings, his contributions in these fields being notable for a sound admixture

of positivism with Jungian humanistic aspiration.

To quote again from the *British Medical Journal*: '... the younger generations who have profited from his writings and from the unselfish help he has always given to colleagues and students will acclaim him as a great doctor, a wise philosopher, a most courteous gentleman'. His friends will hope that more and happy years remain to him. As he himself put it when he was eighty, likening his life to a strenuous Alpine walking tour, 'This holiday is not yet over'.

E. P. W. HELPS

REFERENCES

1. *British Medical Journal* (1953) 'Dr. F. Parkes Weber, 90.' Editorial, i, 1044.
2. Weber, F. P. (1907) 'Multiple hereditary developmental angiomas (telangiectases) of the skin and mucous membranes associated with recurring haemorrhages.' *Lancet*, ii, 160.
3. — (1918) 'Haemangiectatic hypertrophy of limbs-congenital phlebarteriectasis and so-called congenital varicose veins.' *Brit. J. Child Dis.*, 15, 13.
4. — (1922) 'Right-sided hemi-hypertrophy resulting from right-sided congenital spastic hemiplegia with a morbid condition of the left side of the brain, revealed by radiograms.' *J. Neurol. Psychopath.*, 3, 134.
5. — (1925) 'A case of relapsing non-suppurative nodular panniculitis showing phagocytosis of subcutaneous fat cells by macrophages.' *Brit. J. Derm.*, 37, 301.
6. — Watson, J. H. (1904) 'A case of chronic polycythaemia with enlarged spleen, probably a disease of the bone marrow.' *Trans. clin. Soc. Lond.*, 37, 115.
7. — (1908) 'Arteritis obliterans of the lower extremity with intermittent claudication. ('Angina cruris').' *Lancet*, i, 152.
8. Buerger, L. (1908) 'Thrombo-angiitis obliterans: a study of the vascular lesions leading to presenile spontaneous gangrene.' *Amer. J. med. Sci.*, 136, 567.
9. Weber, F. P. (1897) 'A case of localised sweating and blushing on eating.' *Trans. clin. Soc., Lond.*, 31, 277.
10. — (1926) 'Cutaneous striae, purpura, high blood-pressure, amenorrhoea and obesity.' *Brit. J. Derm.*, 38, 1.
11. Cushing, H. (1932) 'The basophil adenomas of the pituitary body and their clinical manifestations (pituitary basophilism).' *Bull. Johns Hopk. Hosp.*, 50, 137.
12. Weber, F. P. (1933) 'Congenital deficiency of conscious auditory perception of words (word deafness) with remarks on other deficiencies of conscious perception.' *J. Neurol. Psychopath.*, 13, 344.
13. — (1942) 'Agnosia of hemiplegia and of blindness after cerebral embolism.' *Lancet*, i, 44.
14. — (1944) 'Explanation of the urge to collect.' *Med. Press*, 211, 414.
15. Paget, J. (1882) 'The Bradshawe Lecture on some rare and new diseases.' *Lancet*, ii, 1017.
16. Bashford, H. (1951) 'A century of physicians.' *Brit. med. J.*, i, 636.

EDITORIALS

Animal Behaviour

THE WORK OF HARLOW

A RECENT series of television programmes has stimulated not only lay but also medical interest in the behaviour and needs of infant mammals. Harlow and his associates at the University of Wisconsin have been attempting to study what they call the strong attachment of the infant animal to its mother. They are interested in getting experimental evidence with regards to the factors making this bond and, particularly, in deciding between an explanation in terms of learning and one based on the concept of innate need or instinct. In an experiment using infant Macaque monkeys they tried to parcel out what they thought were the various components giving rise to the affectional tie. They concentrated mainly on two of them—feeding and the comfort of contact. The monkeys in question were raised without any actual contact with their mothers. Instead they were placed in cages which contained wire frames into which a nursing bottle could be placed. Some of the frames had terry-cloth covers on them and a stylised sort of head. These were called 'inanimate surrogate mothers'. The constructions rested on platforms on which there was room for the baby monkey to lie. Measurements were made of the amount of time the monkeys spent daily lying on the cloth construction and lying on the wire one

when they had equal access to each. Whether they were fed on the wire or on the cloth construction they spent very little time lying on the wire one, both groups equally preferring contact with the cloth 'inanimate surrogate mother'. No matter whether they were fed on a cloth or wire construction, they all reacted identically to a novel or disturbing experience, such as putting a moving toy band into their cage; they would immediately run to the cloth construction and rub their bodies on it. After an interval they would look at the novel object, some of them leaving the platform and going towards it.

Some infant monkeys were raised with only a wire frame feeding construction, while others were raised in a cage with only a cloth feeding construction. Those who had the wire frame did not cling to or embrace it as did those who had the cloth 'mother'. The infants without the cloth 'mother' would clutch themselves, rock, make sounds and suck more than the others and the authors felt this was 'greater emotionality in the presence of fear-provoking stimuli'. Infant monkeys who were placed in a strange environment after having been brought up in the manner described reacted differently. Those who had been brought up with only the wire construction showed a far greater emotional response than those who had been

raised with the cloth construction. Those raised with the wire construction engaged in much less exploratory and manipulative activities. Typical responses were freezing in a crouched position, or running round the room on their hind legs and clutching themselves with their arms. Testing the monkeys by putting them in a box where they could see but not touch either the cloth or wire mother construction, another monkey or an empty box respectively, demonstrated that most lever pressing was done in response to the sight of another monkey. But those raised with cloth or cloth and wire showed almost the same amount of lever pressing when they got a view of the cloth mother construction. These trends continued even after the various contraptions were permanently removed from their cages.

This series of experiments, which has here been only very briefly described, indicates that a source of extensive, soft body contact can be an important variable in determining the speed and relative adequacy with which novel disturbing environmental stimuli problem situations can be mastered. All of

this has, of course, many implications with regard to the actual physical relationship between human mothers and their children. Physical contact between the mother and the child while the child is feeding, the physical presence of the mother while the child is introduced to novel situations, and even sight of the mother all seem to be powerful influencing factors which help to determine the adequacy with which a child explores his environment and meets novel and disturbing situations.

The study by HARLOW and his associates is an attempt, by means of laboratory experiments, to tease out and examine one element of the complexity of mother child relationships. Even the element which they have studied appears to be a highly complex thing.

Mr. S. A. BARNETT drew attention to various aspects of animal behaviour and their relation to medicine in a recent article in *The Lancet* (1961). The same author is preparing a booklet on animal behaviour which will shortly be published as a *Little Club Clinic in Developmental Medicine*.

MARTIN BAX

Changes in the Circulation at Birth

IN the recent National Survey of Perinatal Mortality,¹² in England, Scotland and Wales the 'respiratory distress syndrome' was the commonest cause of death in newborn infants during the first week. Hyaline membrane or massive pulmonary haemorrhage was present in 22 per cent of infants (197 of 901). 'Hyaline membrane disease' is responsible for about a third of the neonatal deaths in the United States¹.

The numerous hypotheses suggested to account for this disease include increased surface tension of the alveoli³, decreased pulmonary fibrinolytic activity¹¹, vasomotor hypotonia^{14, 15, 16}, and injury to pulmonary capillaries^{5, 8}. The concept that the respiratory distress syndrome resulted from failure of circulatory adjustments at birth was stressed by LENDRUM¹⁰ and BONHAM CARTER⁶. Some support for this hypothesis has come from cardiac catheterisation studies of newborn infants¹⁸. These studies showed that infants with severe respiratory distress have a widely patent ductus with a large left-to-right shunt and in some instances a right-to-left shunt. Their pulmonary and systemic arterial pressures are lower than in normal infants. Circulatory changes immediately after birth are therefore of great interest to paediatricians and obstetricians, but only a few advances in this field have come from the study of human infants. Most of our knowledge has been obtained by physiologists working on other mammals.

Dr. GEOFFREY DAWES, who has contributed so much to our understanding of the foetal and newborn circulations, has lately reviewed our present knowledge in the *British Medical Bulletin* (1961, 17, 148). One of the most striking concepts raised by DAWES and his associates is that the circulation of neonates differs both from that of the foetus and that of the adult. In the newborn the ductus and the foramen ovale gradually close and there are large changes in the pulmonary and systemic vascular resistances, pressures and flows. The magnitude of circulatory adjustments during this critical period will be apparent when one considers that at the time of delivery more than half of the infant's combined ventricular output is passing through the placenta. When the umbilical cord is tied the systemic vascular resistance rises sharply. After expansion of the lungs there is a fall in the pulmonary vascular resistance and pulmonary arterial pressure, a large and abrupt increase in the pulmonary blood-flow, and a rise in the left atrial pressure.

In the (sheep) foetus the de-oxygenated blood from the superior vena cava passes completely into the right ventricle, but only about half of the more oxygenated blood from the inferior vena cava containing the placental venous return passes into the right ventricle; the remainder streams into the left atrium through the foramen ovale. The blood in the carotid arteries

comes entirely from the left ventricle and has a higher oxygen saturation than that in the descending aorta, which also receives blood from the pulmonary artery through the ductus. The ciné-angiographic studies of LIND and WEGELIUS¹² suggest that the human foetus has a similar circulation.

Contrary to former belief, the main autonomic reflexes function at birth; thus, the Hering-Breuer, carotid body chemoreceptor, pulmonary respiratory chemoreceptor, and baroreceptor reflexes have been demonstrated in newborn rabbits. DAWES argues that, since these reflexes are active, the stimulus for the onset of breathing is probably a fall in arterial oxygen saturation. The mean arterial oxygen saturation in the descending aorta of foetal rhesus monkeys was 58 per cent, but that in the umbilical artery after delivery was 30 per cent. This fall in arterial oxygen content during delivery is attributed to compression of the umbilical cord, partial separation of the placenta, and a reduction in uterine blood-flow during contractions.

The ductus arteriosus does not close at once at birth either in man^{2, 9, 15} or in lambs, but in lambs it constricts after ten to thirty minutes of adequate pulmonary ventilation. The flow through the ductus reverses because of an increase in systemic and a decrease in pulmonary vascular resistance. Transient murmurs, probably arising from the patent ductus, have been heard in lambs by DAWES and in humans by others^{4, 7}. The ductus is closed by contraction of muscle in its wall, the main stimulus to its closure seeming to be an increase in the arterial oxygen

saturation, although other factors contribute. DAWES has demonstrated that artificially induced hypoxia may lead to reopening of the ductus but only during the first five hours of life. When the ductus is artificially closed and the left-to-right shunt thus obliterated, the pulmonary artery flow diminishes, the pulmonary vascular resistance rises, and the pressures in the pulmonary artery and left atrium fall. The pulmonary artery pressure, however, continues to fall over a period of weeks after the ductus has closed, probably because of changes in the calibre and structure of the pulmonary vessels.

The foramen ovale closes when the pressure in the left atrium exceeds that in the inferior vena cava and in the right atrium. With the onset of pulmonary ventilation and increase of blood-flow through the lungs, the pressure in the left atrium rises and comes to exceed the pressure in the inferior vena cava, which falls after the umbilical vein is ligated because the large venous return from the placenta is cut off from the inferior vena cava. Ductus closure, however, tends to have the opposite effect—it diminishes the left atrial pressure more than it does the pressure in the inferior vena cava. In kittens it has been shown that there is a gradual increase in pressure gradient between the left and right atria. Thus, at three days only a slight gradient is present while at two months the left atrial pressure is almost double the right. The time of closure of the foramen ovale differs in various mammals, occurring within a few hours in foals, at the end of the first week in lambs and after twelve days in rhesus monkeys.

The oxygen consumption in the lamb per unit of body-weight remains unchanged during the last third of gestation but there is almost a threefold increase in consumption within twenty-four hours of birth; a similar but smaller increase occurs in other mammals and probably also in humans. It is of interest that hypoxia increases the cardiac output from three to five times in adult sheep but not at all in newborn lambs. DAWES suggests that this is

because the cardiac output of the newborn may already be maximal.

There is still a long way to go before the advances in foetal and neonatal physiology of experimental animals can be applied to the clinical problems of newborn human infants. It is hoped that further investigations of circulatory changes both in the animal laboratory and in the wards may help to bridge this gap.

P. F. BENSON

REFERENCES

1. Aronson, N. (1961) 'Studies on hyaline membranes.' *Pediatrics*, **27**, 567.
2. Adams, F. H., Lind, J. (1957) 'Physiologic studies on the cardiovascular status of normal newborn infants (with special reference to the ductus arteriosus).' *Ibid.*, **19**, 431.
3. Avery, M. E., Mead, J. (1959) 'Surface properties in relation to atelectasis and hyaline membrane disease.' *Amer. J. Dis. Child.*, **97**, 517.
4. Benson, P. F., Bonham Carter, R. E., Smellie, J. M. (1961) 'Transient and intermittent systolic murmurs in newborn infants.' *Lancet*, **i**, 627.
5. Berfenstam, R., Edlund, T., Zettergren, L. (1958) 'The hyaline membrane disease; a review of earlier clinical and experimental findings and some studies on the pathogenesis of hyaline membrane in O_2 -intoxicated rabbits. *Acta Paediatr., Uppsala*, **47**, 82.
6. Bonham Carter, R. E. (1957) 'The architectural function of pulmonary capillaries.' *Lancet*, **i**, 1292.
7. Burnard, E. D. (1958) 'A murmur from the ductus arteriosus in the newborn baby.' *Brit. med. J.*, **i**, 806.
8. Ingalls, T. H. (1954) 'Epidemiology of retrolental fibroplasia: its etiologic relation to pulmonary hyaline membrane.' *New Engl. J. Med.*, **251**, 1017.
9. James, L. S., Rowe, R. D. (1957) 'The pattern of response of pulmonary and systemic pressures in newborn and older infants to short periods of hypoxia.' *J. Pediatr.*, **51**, 5.
10. Lendrum, F. C. (1955) 'The "pulmonary hyaline membrane" as a manifestation of heart failure in the newborn infant.' *Ibid.*, **47**, 149.
11. Lieberman, J. (1959) 'Clinical syndromes associated with deficient lung fibrolytic activity. I. A new concept of hyaline-membrane disease.' *New Engl. J. Med.*, **260**, 619.
12. Lind, J., Wegelius, C. (1954) 'Human foetal circulation: changes in the cardiovascular system at birth and disturbances in the post-natal closure of the foramen ovale and ductus arteriosus.' *Cold Spr. Harb. Symp. quant. Biol.*, **19**, 109.
13. 'National survey of perinatal mortality.' (1961) *Brit. med. J.*, **i**, 1313.
14. Neligan, G. A. (1959) 'The systolic blood-pressure in neonatal asphyxia and the respiratory distress syndrome.' *Amer. J. Dis. Child.*, **98**, 460.
15. Rudolph, A. M., Auld, P. A. M., Drorbaugh, J. E., Rudolph, A. J., Nadas, A. S., Smith, C. A., (1959) 'Studies on the circulation of infants with hyaline membrane disease.' (Abstract) *Ibid.*, **98**, 630.
16. Rudolph, A. M., Drorbaugh, J. E., Auld, P. A. M., Rudolph, A. J., Nadas, A. S., Smith, C. A., Hubbell, J. P., (1961) 'Studies on the circulation in the neonatal period. The circulation in the respiratory distress syndrome.' *Pediatrics*, **27**, 551.

The Principles of Hughlings Jackson Applied to Psychiatry

Henri Ey's Dynamic Concept of Neuro-Psychiatry

The ideas of Hughlings Jackson have decisively influenced the neurological thinking of the last hundred years. Their impact on psychiatry, however, has been negligible, and the organo-dynamic concept of Henri Ey, in Paris, is the first major and systematic attempt to apply them to psychological disorders. The following is a brief account of this important and stimulating approach. It was intended to publish it at the time of the anniversary of Hughlings Jackson's death, but translation difficulties delayed publication.

NEUROLOGY and psychiatry are complementary disciplines: neurology deals with the partial disintegration of sensorimotor functions, while psychiatry is the pathology of integrated nervous activity at its highest level.

The organo-dynamic concept of psychiatry postulates a two-dimensional organisation of mental activity, one developmental and the other cross-sectional or structural. From infancy to adult life, human mental activity passes through a series of developmental stages to achieve an organisation which, by its very nature, is never entirely finished. This is the basis of the concept of evolution in human psychology. Psychic life, thus differentiated and integrated in the service of man's adaptation to his environment, can also disintegrate (destruction of consciousness). The dream epitomises this: the higher level of psychic organisation prevalent during waking hours is swept away—disintegrates—during sleep and psychic life functions at the lower level of dreaming. Psychology and psychopathology suppose therefore the dimension

of psychic evolution and the dimension of a hierarchy of integrative levels corresponding to the different stages of this evolution.

This organo-dynamic concept of psychiatry rests on four fundamental principles:

(1) This is an application of the Jacksonian idea that *disease does not create but releases*. The different aspects of psychic life, called intelligence, affectivity and personality, constitute higher levels of organisation; their dissolution, or 'destruction', releases lower or archaic levels which represent the various forms and degrees of mental disease; this is analogous to the Freudian notion of regression. Mental disease is therefore contained in the hierarchic organisation of one's psychic being. Potentially everyone is mentally ill; he actually becomes mentally disordered when falling asleep, and still more so when the structure of his psychic being is disorganised in a lasting manner.

(2) *The structure of mental disease is essentially negative*—i.e., an application of the Jacksonian principle of the negative or defect character of neurological disease. This negative aspect is readily seen in dementia, and the concept of 'immaturity' in the field of neuroses is of the same order. The existential analysis of mental diseases has given new insights revealing this defect character of morbid psychological experience.

(3) *The forms of mental disease represent different levels of dissolution of psychic organisation*. Applying the concept of levels of dissolution formulated by Hughlings Jackson, it is possible to classify mental diseases according to the hierarchic organisation of psychic life. Seen in this light mental disease is a group of symptoms corresponding to a certain level of dissolution of the psyche, taking a definite and clinically identical course.

1, Ey, H. and Rouart, J. 'Essais d'application des principes de Jackson à une conception dynamique de la neuro-psychiatrie'. *Encephale*, 1936, 1, 313-356; 2, 30-60 and 96-123.

Ey, H. *Études Psychiatriques*, 1952-1961. Paris: Desclée de Brouwer.

The classification of mental diseases put forward by Henri Ey is based on the principle of the two-dimensional organisation of psychic life—the dimension of the actual field of consciousness and the dimension of the historical development of personality—as follows:

Pathology of Consciousness <i>(Acute Psychoses)</i>	Pathology of Personality <i>(Chronic Psychoses and Neuroses)</i>
Manic-depressive attacks Delirious, hallucinatory and dream states Oneiroid psychoses	Neuroses and personality disorders Chronic delirium and schizophrenia Dementias

The field of consciousness consists of the pattern of current perception; organised on functional levels, it is responsible for the apparent order of our temporal and spatial experience. All acute psychoses—manic-depressive, epileptic, oneiroid and delirious—present clinically as levels of destruction of the field of consciousness, analogous to what happens in sleeping and dreaming.

The personality system—i.e., the permanent structure of the ego—has a development, an organisation and a disorganisation of its own. In dementia, schizophrenia, paranoia and neurosis the patient builds up a personal system and functions at this morbid and 'disorganised' level. By the very fact that such diseases acquire the structure of a permanent personality system they have a necessary aspect of chronicity.

(4) *Mental diseases are organically determined:*
the concept of the disorganisation of the whole

structure of the psyche must become the basis of psychiatric aetiology. The clinical pictures characteristic of various mental diseases result from the combined efforts of dissolution at the higher level (negative factor) and liberation of lower level activities (positive factor). The dissolution is an organic process; the manifestations or content of the disease are determined by psychological forces. Mental disease is an impoverishment of human existence, a 'pathology of release'.

This organo-dynamic concept of psychiatry has received some support from recent developments in neurophysiology and psychopharmacology. Increasing knowledge about the role of the centrencephalic system and of the rhinencephalon suggest that the functional disintegrations of this vast system are related to the states of delirium, epilepsy and manic-depressive psychosis. The model psychoses produced by some hallucinogenic drugs can be regarded as experimentally produced dissolutions of consciousness.

It is also claimed that such a concept avoids the oversimplifications of psychogenesis (causation by more or less unconscious psychological intention) and sociogenesis (causation by environmental stresses), as well as the absurdities of mechanistic theories—i.e., causation by localised cerebral lesions.

A. G. MEZEY

Red-cell Enzymes and Severe Neonatal Jaundice

In the last few years, reports from Italy, Japan, Singapore, Greece and South Africa¹⁻⁴ have pointed out that in these countries a substantial number of the cases of severe neonatal jaundice or kernicterus in full-term infants are not due to isoimmunisation (Rhesus disorder). A familial incidence was noted by MORGANTI and BEOLCHINI^{1, 7} and the Greek workers were impressed by both the haemolytic and the familial character of the jaundice. A preliminary survey done by these Greek workers excluded any connection between neonatal jaundice and the haemoglobinopathies prevalent in the Mediterranean area. Next they examined the connection with another familial haemolytic mechanism—that of favism. Favism is a very old disease in some areas but its mechanism was only recently elucidated by work done on primaquine-sensitive negroes. The American Malaria Research Unit⁸ localised the defect of the drug-sensitive subjects in the erythrocytes, and it was subsequently proved that a deficiency of the red-cell enzyme glucose-6-phosphate dehydrogenase (G-6-P.D.), was the basis for the development of haemolysis on exposure to primaquine and some other drugs or to fava beans.^{9, 10}

The development by MOTULSKY and CAMPBELL¹¹ of an easy and reliable test for assessing the activity of this enzyme made it possible to screen a large number of persons. Thus DOXIADIS, FESSAS and VALAES^{12, 13, 14} were able to

present conclusive evidence of an aetiological connection between G-6-P.D. deficiency and the development of severe neonatal jaundice; in a control group of 500 male infants only 3 per cent showed G-6-P.D. deficiency, compared with 77 per cent of the infants with severe jaundice without isoimmunisation. Meanwhile cases of severe jaundice or kernicterus in infants with G-6-P.D. deficiency have been reported from Sardinia and Singapore.^{15, 16, 17}

The group of full-term infants with severe neonatal jaundice not due to isoimmunisation is numerically very important in some areas. In the Greek population a third of all the cases treated by exchange transfusion belonged to this group. Among the infants admitted with established kernicterus 80 per cent belonged to the group without incompatibility. Severe neonatal jaundice due to G-6-P.D. deficiency is a much more difficult problem for the practitioner than the jaundice due to isoimmunisation. Icterus seldom appears in the first twenty-four hours of life so that its pathological nature is not apparent from the beginning. In some cases jaundice starts as late as the fourth day of life, while the severity of the hyperbilirubinaemia is independent of the day of the appearance of jaundice. Rising bilirubin values and the development of kernicterus has been seen even after the 10th day of life.¹⁸ Kernicterus never appears so late in cases of haemolytic disease due to

isoimmunisation. The unusual and unpredictable course explains why practitioners often failed to refer their cases for treatment in time. Exchange transfusion is as effective in this group as in cases of isoimmunisation. For the indications for this treatment one has to rely solely on the bilirubin level. The detection of the enzyme defect does not simplify the indications for treatment. Only a small proportion of the infants with the defect exhibit jaundice severe enough to require protection by exchange transfusion.

In some cases an extrinsic haemolytic factor—the inhalation of naphthalene or the administration of vitamin-K analogues—seemed to aggravate or precipitate the haemolysis, but in most cases no such extrinsic factor could be detected. Since spontaneous haemolysis is clearly confined to the neonatal period, we must accept that one or more of the components of the newborn's unstable 'milieu interieur' must provide the noxious mechanism.

As a rule, the haemolysis seems to affect only a small proportion of the red-cell population, high serum-bilirubin values occur for only a short time, and no overt anaemia or morphological abnormalities are seen. In other cases the destruction affects almost all the red cells. Anaemia with haemoglobin values of 4 to 5 g. per 100 ml. occurs in a matter of a week, while the peripheral blood shows extreme fragmentation of the red cells and Heinz body formation.^{14, 18} In the natural course of the disease, without any interference by exchange transfusion, these cases show both a protracted hyperbilirubinaemia and high conjugated bilirubin values.

Haemolysis is probably preceded by morphological abnormalities in all cases, but in the group with self-limited haemolysis this stage is missed since the first examination is not done until after these damaged cells have been removed from the circulation.

With the demonstration that G-6-P.D. deficiency can cause severe neonatal jaundice a new cause of neonatal haemolysis has emerged. This is the first instance in which excessive rates of haemolysis in the newborn have been attributed to a disturbance of the red-cell metabolic pathways. Another important point is that a defect which in later life might remain completely latent is dramatically manifested in the neonatal period. It is quite likely that there are other red cell enzyme defects, hereditary or even transient, causing haemolysis and icterus in the neonatal period. The Greek group have already collected a number of cases where both familial incidence and evidence of haemolysis point to an enzymatic mechanism, though in these cases a deficiency of G-6-P.D. has been excluded.¹⁴ The 'infantile pyknocytosis' described by TUFFY and others,¹⁹ as well as the cases described by GASSER and KARRER,²⁰ ALLISON,²¹ and VARADI, and HURWORTH,²² all of which are characterised by morphological changes in the red cells and/or the appearance of Heinz bodies *in vivo*, may well be examples of haemolysis caused by a disturbed red-cell metabolism, due to (a) a hereditary enzyme defect, (b) a transient instability, or (c) interference with intracellular metabolism by toxic vitamin-K analogues. Cases of severe jaundice in full-term infants which have

hitherto been attributed solely to hepatic insufficiency should be re-examined in the light of this new concept.

The cases of severe jaundice due to enzyme deficiency are unique in another respect—namely, that rising and very high bilirubin values sometimes occur in these cases after the first week of life.

The occurrence of kernicterus in such cases on the 10th day of life and even later shows that one should not rely on the maturation of the 'blood-brain barrier' to protect the nerve cells from the toxic effects of high bilirubin concentrations.

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REFERENCES

1. Morganti, G., Beolchini, P. E. (1950) 'The existence and frequency of cases of familial icterus in the newborn without serological indications of maternal-foetal immunisation.' (In Italian.) Proceedings of the 1st Congress of Nephrology, Triest.
2. Sutow, W. W., Moloney, W. C., Margoles, C. (1956) 'Kernicterus in Japanese infants. 1. Clinical and serological data from 25 patients.' *Pediatrics*, **17**, 349-358.
3. Wong, H. B. (1957) 'Kernicterus not associated with haemolytic disease.' *Arch. Dis. Childh.*, **32**, 85-90.
4. Maycock, H. G. T., Gibson-Hill, M. M. H. (1954) 'Haemolytic disease of the newborn in Singapore.' *Med. J. Malaya*, **8**, 343-350.
5. Doxiadis, S. A., Paraskevopoulos, H. (1959) 'The prevention of kernicterus.' *Proc. med. Soc. Athens*.
6. Shnier, M. H., Levin, S. E. (1959) 'Hyperbilirubinaemia and kernicterus in premature and full-term Bantu newborn infants.' *Brit. med. J.*, **1**, 1004-1007.
7. Morganti, G., Beolchini, P. E. (1954) 'New data on familial icterus of the newborn.' (In Italian.) *Minerva pediatrica*, **Torino**, **6**, 202-205.
8. Beutler, E. (1959) 'The hemolytic effect of primaquine and related compounds: a review.' *Blood*, **14**, 103-139.
9. Sansone, G., Segni, G. (1956) 'First determinations of glutathione (GSH) in the blood in favism.' (In Italian.) *Boll. Soc. ital. Biol. sper.*, **32**, 456-458.
10. Szeinberg, A., Sheba, C., Hirshorn, N., Bodonyi, E. (1957) 'Studies on erythrocytes in cases with past history of favism and drug-induced acute hemolytic anemia.' *Blood*, **12**, 603-613.
11. See note at end of this article.
12. Doxiadis, S. A., Fessas, Ph., Valaes, T. (1960) 'Erythrocyte enzyme deficiency in unexplained kernicterus.' *Lancet*, **ii**, 44-45.
13. Doxiadis, S. A., Fessas, Ph., Valaes, T., Mastrokalos, N. (1961) 'Glucose-6-phosphate dehydrogenase deficiency; a new aetiological factor of severe neonatal jaundice.' *Ibid.*, **i**, 297-301.
14. Fessas, Ph., Valaes, T., Doxiadis, S. A. (1961) 'Study of new aetiological factors of neonatal jaundice.' *Proc. med. Soc. Athens*.
15. Panizon, F. (1960) 'Severe icterus of the newborn associated with a deficiency in glucose-6-phosphate dehydrogenase.' (In French.) *Biol. neonat.*, **2**, 167-177.
16. Smith, G. D., Vella, F. (1960) 'Erythrocyte enzyme deficiency in unexplained kernicterus.' *Lancet*, **i**, 1133-1134.
17. Weatherall, D. J. (1960) 'Enzyme deficiency in haemolytic disease of the newborn.' *Ibid.*, **ii**, 835-837.
18. Valaes, T., Fessas, Ph., Doxiadis, S. A. (1961) 'Kernicterus in full-term infants without isoimmunisation (4 cases).' *Proc. roy. Soc. Med.*, **54**, 331-333.
19. Tuffy, P., Brown, A. K., Zuelzer, W. W. (1959) 'Infantile pyknocytosis.' *A.M.A. J. Dis. Child.*, **98**, 227-241.
20. Gasser, C., Karrer, J. (1948) 'Spontan "Innenkörperbildung" mit deletärer hämolytischer Anämie bei Frühgeburt.' *Schweiz. med. Wschr.*, **78**, 974-975.
21. Allison, A. C. (1957) 'Acute haemolytic anaemia with distortion and fragmentation of erythrocytes in children.' *Brit. J. Haematol.*, **3**, 1-18.
22. Varadi, S., Hurworth, E. (1957) 'Heinz-body anaemia in the newborn.' *Brit. med. J.*, **i**, 315-318.

Motulsky and Campbell Method for Estimating G-6-P.D. Activity of Red Cells

This method has not apparently been published by its authors, but the following brief description is taken from a paper on 'Glucose-6-phosphate dehydrogenase deficiency', by S. A. Doxiadis, Ph. Fessas, T. Valaes and N. Mastrokalos in *The Lancet* (1961, **1**, 297), by kind permission of the Editor.

0.01 ml. of packed red cells is added to 1.0 ml. of distilled water and 0.2 ml. of tris-buffer, 0.74 M, pH 8.5. To this are added in succession:

0.05 ml. of a solution of 0.165 g. glucose-6-phosphate, disodium salt, in 10 ml. of water ('Sigma').

0.05 ml. of a 0.1 per cent solution of triphosphopyridine nucleotide (British Drug Houses).

0.25 ml. of a solution of 0.32 g. brilliant cresyl-blue in 1000 ml. of water (National Aniline).

After good mixing, the reagents are overlaid with mineral oil. The tubes are then incubated in a water bath at 37° C. and inspected for decolourisation against daylight at 50, 75, 100, 120, 180 minutes, and 6 hours.

All samples of blood where decolourisation was complete at 100 minutes were considered as exhibiting normal G-6-P.D. activity (non-reactors). All samples, either from cord blood or from adults, which were not decolourised at all up to 3 or 6 hours were considered as having practically no enzyme activity (reactors). Partial decolourisation at 100 minutes was taken as a sign of partial enzyme deficiency (intermediate reactors).

NOTICE

International Conference on the Backward Child

London: April 16-19, 1962

THE Guild of Teachers of Backward Children are running this conference at Goldsmiths College, University of London, to consider the following challenging themes:

- (a) The content of present provision (with emphasis on different approaches and trends).
- (b) What the teacher can learn from other disciplines (e.g., from work with the deaf or other handicapped children).
- (c) What research suggests for future practice.

These three topics will be considered in joint session, but in addition the conference will break up into smaller groups to examine the following:

- (a) The Primary School
- (b) The Secondary School
- (c) The Special (ESN) School
- (d) The Residential (ESN) School
- (e) The Training Centre

Dr. W. D. Wall, Director of the National Foundation for Educational Research, and Dr. N. O'Connor, of the Medical Research Council Unit, Maudsley Hospital, will be among the speakers. A programme of visits is being arranged to special schools, remedial departments in secondary schools, training centres and institutions. In addition several exhibitions are being arranged, including one by the National Book League, 'The Teacher and the Backward Reader'. There will also be films.

Those who wish to attend should contact the ticket secretary, Miss P. Burnell, 125 High Holborn, London, W.C.1. Those wishing to contribute to any of the sessions should write to the conference organiser, Mr. S. S. Segal, J.P., at the same address. Hostel accommodation is available but early booking should be made.

Martin Bax

When is a Child with Cerebral Palsy Ineducable?

THE term 'ineducable' is applied by educationists to children whose intelligence is so limited that they are incapable of formal learning in the basic school subjects. Most of those responsible for advising on provisions for the teaching of children of limited intelligence do not like to draw a sharp line of demarcation between children who need special school provision and the 'ineducable', because various other factors as well as mental subnormality must be considered before deciding that a child is ineducable. Moreover, when children are deemed ineducable they are removed from the responsibility of an Education Authority, and the flexibility of movement and ease with which a child can be re-admitted to an Education Authority school varies from one Local Authority to another.

Experience suggests that a non-handicapped child whose intelligence quotient is below 55 on a test such as the Stanford-Binet, provided of course that the test conditions make it a reasonably valid estimate, is likely to prove incapable of receiving education at school and would be more suitably catered for in a Training Centre. It is the Local Health Authority's duty to provide such centres for these children, but the adequacy of the provision still varies considerably* and the quality of the centres themselves is also very uneven. Where there are good Training Centres and Medical Officers whose

interest embraces these children as well as those in special schools for subnormal children within the school system, movement between these two types of provision can remain flexible enough to gather the fruits of observation and experiment without much harm being done to the child who has been wrongly assessed.

The 'other factors' that should be taken into account when estimating the educability of children of borderline capacity include family background, emotional stability, relevant personality traits, social adjustment, and capacity to fit into a group without seriously disrupting the other children. It is not normally the practice to make a hard decision on such borderline cases until they have had a trial period in a special school for educationally subnormal children.

Because of their multiple handicaps, the decision regarding cerebral palsied children is far more difficult, since their handicaps vary widely both in range

* According to 'Community Care of the Mentally Handicapped', published by the National Association for Mental Health in 1960, the following Local Authorities then provided 60 or more places in Training Centres per 100,000 population: Barnsley, Bradford, Dewsbury, Dudley, Exeter, Great Yarmouth, Ipswich, Kingston-upon-Hull, Leeds, Lincoln, Oldham, Rotherham, Stoke-on-Trent, Wolverhampton, York, Cardiff, Swansea and Isle of Ely. The following provided less than 10 places per 100,000: Cornwall, Hunts, Rutland, Suffolk West, Sussex East, Yorks (East Riding), Anglesey, Brecon, Caernarvon, Cardigan, Merioneth, Montgomery, Pembroke, Radnor, Isles of Scilly, Barrow, Worcester and Merthyr Tydfil.

and severity. As well as mental and motor handicap, many cerebral palsied children suffer also from defects of vision, hearing, sensation and speech, and perhaps from epilepsy as well. Sensory defects, and lack of normal sensori-motor experiences owing to unavoidable environmental restrictions, must contribute to the difficulty of making a true measure of a cerebral palsied child's potential for learning. Such children learn more slowly than their normal fellows and require more time to consolidate what they have learnt. Inevitably, also, more of their time and energy is spent on other necessary forms of training, such as motor or speech training. The cerebral palsied child therefore requires an assessment period of at least a year before a final decision regarding educability is reached, particularly if his physical handicaps are severe and if difficulties in communication make it impossible for him to demonstrate his capacity for understanding through his response to ordinary tests. During this assessment period any therapy that seems likely to help the child should be available to him. But the assessment centre's staff should also include teachers trained and equipped to catch the moment of the child's readiness for ventures into learning by extending his skills and experience through play and experimentation. Unfortunately, most training or occupation centres do not yet provide this kind of highly skilled staff.

Educability, in its strict meaning of capacity to profit by formal teaching in the basic subjects of reading, writing, spelling and arithmetic, depends

primarily on the child's inherent intelligence. The Education Act sets the lower limit for an unhandicapped child at an I.Q. in the fifties, but educationists' opinions vary regarding the intellectual requirements for cerebral palsy children. In her survey of 1952, DUNSDON¹ found that among cerebral palsied children whose degree and type of difficulty prevented them from attending a school for physically handicapped children, 70 to 80 per cent were of an intellectual dullness which debarred them from benefiting from formal education. She maintains that, during the early school years, attention can be directed more profitably to the social and physical aspects of training than to formal education, and that if social and physical training is begun early enough it should be possible to differentiate, by the time the child is about 9 years old, between those who have made good enough general progress to enable them to profit from formal education and those who are unlikely to do so. By this time, intensive therapy should no longer be required, and children are better able to attempt formal work and make educational progress when their energy is not simultaneously being drained by intensive physical treatment. DUNSDON holds that cerebral palsied children suitable for such transfer would need an I.Q. of at least 85. SCHONELL², however, would place the base line at I.Q. 70, and she urged that physical treatment and formal education should be combined.

These widely divergent views are both concerned with children whose potential intelligence, whatever their physical handicap, can be estimated with reasonable certainty, and who,

given suitable opportunities and teaching, are capable of making some progress in formal learning. Here, however, we are concerned with borderline cases lying in the uncertain no-man's-land between this potentially teachable group and those whose low intelligence leaves no doubt regarding their capacity to benefit from formal education, however rudimentary. For cerebral palsied children, many more factors must be recognised and evaluated before deciding whether they are educable. If there is any doubt, these children should be on trial for at least a year, preferably in an assessment centre, but otherwise in a school for physically handicapped children or with a home teacher. During this trial period three factors should help in assessing the child's potential: (1) the extent to which he can be helped to establish communication with his world; (2) his capacity and inventiveness for circumventing his own physical limitations; and (3) the extent to which his capacity for understanding has increased during the assessment period—a valuable indication of his developmental potential.

While the legislative distinction between educable and ineducable children continues, it would be a useful safeguard to retain even grossly handicapped cerebral palsied children in the 'Educationally Subnormal' category until a long trial—even up to three years—has clearly established that they

are ineducable. Children for whom it is possible to demonstrate, even if only in selected areas of intellectual functioning, a level equal to an I.Q. of 60 would be included in this group. We must not lose sight of these children's essential need for *time*. To quote Dr. MARJORIE WILSON:³ 'Many of these cerebral palsied children are years behind in social and physical experience and may (quite irrespective of their chronological age) need the infantile approach to learning, or may even be at babyhood level. Many of them will progress from this stage and show their true potential when they have been given the chance to start from scratch, in spite of their age.' For a large proportion of these children a Training Centre will undoubtedly be the best provision, but until each Training Centre can be staffed with at least one trained teacher capable of providing suitable learning opportunities for children with a wide variety of defects, it seems wise to retain this trial period. The difficulty of deciding whether a child is 'educable' or not—and much of its concomitant anguish—could be minimised by providing, wherever they are geographically possible, small nursery schools for the cerebral palsied, where there would be an opportunity for assessment under trained nursery school teachers in a more relaxed atmosphere for both parents and children.

JESSIE M. WILLIAMS

REFERENCES

1. Dunsdon, M. I. (1952) *The Educability of Cerebral Palsied Children*. London: Newnes, pp. 142-144.
2. Schonell, F. E. (1956) *Educating Spastic Children*. London: Oliver and Boyd, pp. 113-114.
3. Wilson, M. (1955) *The Placement of Cerebral Palsied Children*. Address given at a one-day conference on 'Cerebral Palsy: the Present Position and Future Possibilities.' London: British Council for the Welfare of Spastics.

ORIGINAL ARTICLES

Clinical and Laboratory Diagnosis of Metachromatic Leucodystrophy

BENGT HAGBERG, PATRICK SOURANDER AND LARS SVENNERHOLM

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LATE infantile metachromatic leucodystrophy is suitably classified as a genetically determined, generalised lipidosis due to the accumulation in various organs of lipid metachromatic substances (Brain and Greenfield 1950, Austin 1958, Hagberg *et*

different parts of the world (Austin 1958, Hagberg *et al.* 1959, Jatzkewitz 1958). It is a remarkable fact that disturbances in the metabolism of sphingolipids, the chemical class to which the sulphatides belong, is also found in other diseases

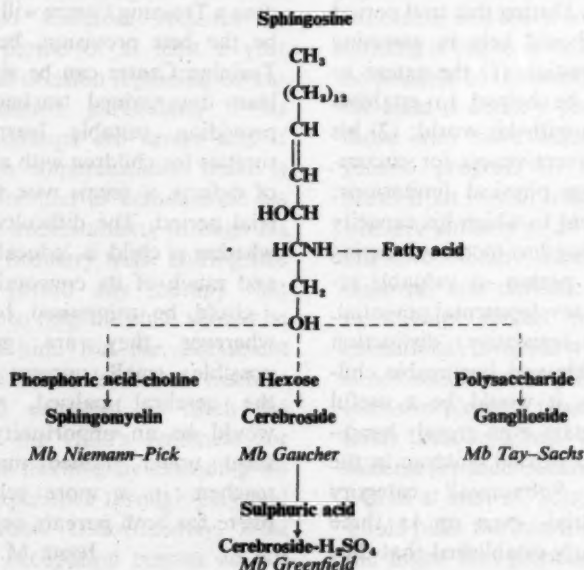


Fig. 1. Diagram showing the close chemical relationship between the neurolipidoses.

al. 1959, 1960). In 1958 these substances were shown to be sulphatides (sulphuric acid esters of cerebroside) by investigations made independently in three

belonging to the neurolipidoses. The close chemical relationship between the various neurolipidoses is schematically shown in Fig. 1. The sulphatides are normal

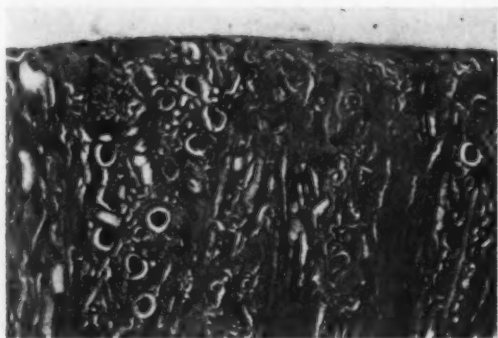


Fig. 2.

Frozen section of kidney stained with cresyl violet acetic acid by v. Hirsch and Peiffer's method, showing abundant red-brown metachromatic deposits in the tubular epithelium.

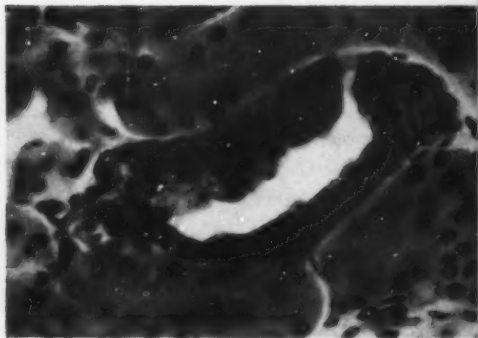


Fig. 3. High power view of deposits.



a b c d e

Fig. 4.

Paper chromatogram showing: (a) 5µg brain sulphatides; (b) urinary lipids from normal child of 5 years; (c) urinary lipids of normal newborn child; (d) urinary lipids of case of metachromatic leucodystrophy; (e) 5µg. brain sulphatides.

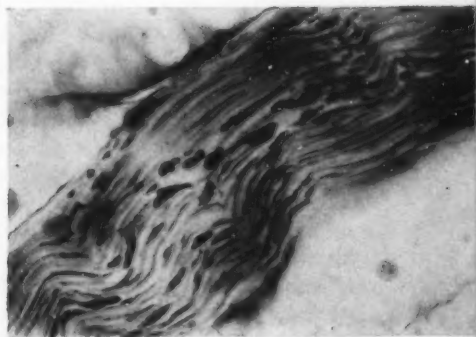
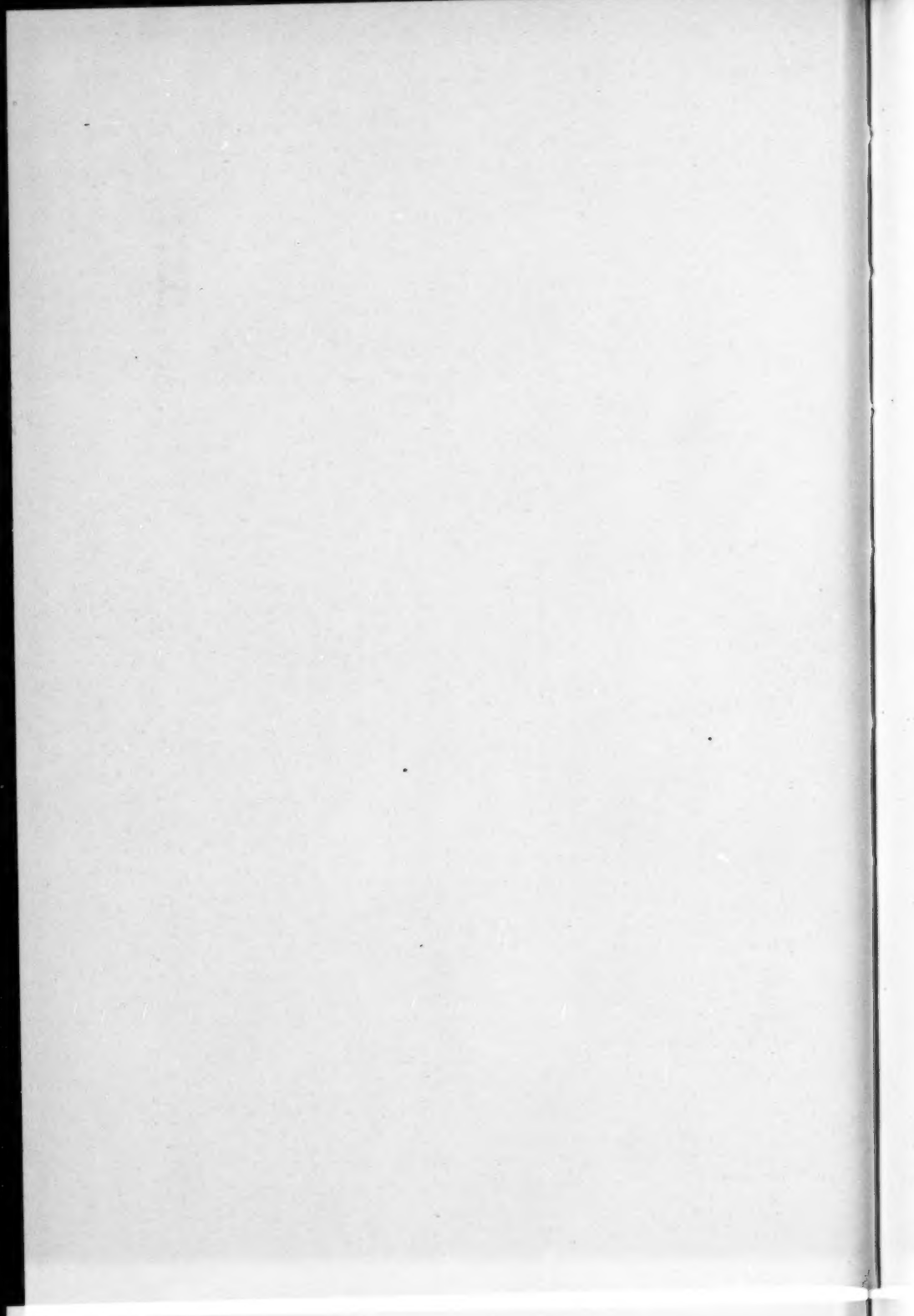


Fig. 5.

Frozen section of sural nerve, stained by the same method as in Figs. 2 and 3. The myelin sheaths are fragmented and partly replaced by large amounts of metachromatic material.



components of the brain lipids and their concentration increases with age.

The enzymatic mechanisms of the normal metabolism of cerebrosides and sulphatides are unknown. Possibly the oligodendroglia cells, which are known to have a high metabolic activity, may be important; and a prominent feature of the present disease is the early disappearance of the interfascicular oligodendroglia (Brain and Greenfield 1950), irrespective of the degree of demyelination.

Clinical symptoms and signs

The neurological picture in this disease is not specific but the symptoms and signs often give valuable clues to the diagnosis. In most cases the first symptoms appear at 1-2 years of age, and are usually observed when the child starts to walk without support. At this early stage the gait is unsteady; the child has knock knees and valgus deformity of the feet; there is general hypotonia but no obvious paresis. Insidiously the child loses first the ability to walk or stand and finally the ability to crawl. The hypotonicity may sometimes be so marked that myopathy may be suspected. The next stage is dominated by flaccid paraplegia or diplegia with diminished or absent tendon reflexes combined with slight signs of pyramidal damage. A second variant is a pure spastic paraplegia with pronounced hypertonus but no signs of secondary motor neurone dysfunction. There is an early increase in the protein content of the cerebrospinal fluid. Spinal tumour, polyradiculitis and cerebral palsy are the common diagnostic errors made at this stage. However, the neurological picture of hypotonia or of spasticity is soon complicated by ataxic signs (coarse tremor, nystagmus and speech disturbances) and at about the same time mental regression and apathy appear, together with peculiar hypertonic fits, cerebral fever and sometimes optic atrophy. Most

patients pass through a complex tetraplegic stage with variable muscle tone ('dystonia' or 'variable rigidity' according to the cerebral palsy nomenclature of some authors). The original dysarthria is converted into a total aphasia. Finally bulbar symptoms complicate the picture, with feeding and respiratory difficulties, and the child usually dies at 3 to 6 years of age from secondary pulmonary infections. In this final stage generalised decerebrate rigidity occurs and cortical blindness and deafness may be added. The child no longer has any contact with his surroundings.

Of large practical, as well as theoretical, interest is the fact that, in this disease, sulphatides also accumulate in organs outside the nervous system. Metachromatic deposits are thus found not only in the nervous system but also in the parenchymal liver cells, in macrophages of the mucous folds of the gall-bladder and in the epithelium of the convoluted and collecting tubules of the kidney. In particular, knowledge of their deposition in the kidney tubules has given us valuable new diagnostic tools, as the substances can be traced in the urine, as was first pointed out by Austin (1957). This tubular background to the urinary findings is illustrated in Figs. 2 and 3, which show large amounts of metachromatic granules in the tubular cells.

Diagnostic Tests on the Urine

The urinary diagnostic tests are of two different kinds: (1) demonstration of metachromatic granular material by microscopy of urinary sediments stained with various aniline dyes; and (2) direct chemical analysis of the urine for sulphatides.

The *microscopical examination* is made possible by the fact that the abnormal granular material can be separated out by centrifugation and can still be stained metachromatically in the urine. The test

is very simple to do and can be performed by the average clinician without technical assistance. For details of the method the reader is referred to Austin's original work (1957). Toluidine blue O in 2 per cent solution produces a golden-brown or red-brown metachromasia of the granular structures. Austin described six different structures, the most specific evidently being large oval granular bodies of mulberry type. However, we have found small amounts of metachromatic material in the form of free granules and casts in healthy children, especially in infants under 1 year of age.

Direct chemical analysis for sulphatides in the urine has been developed by us as a diagnostic test (Hagberg and Svennerholm 1959). In paper partition chromatography with two different solvents, the metachromatic lipids were shown to run identically with a known sample of brain sulphatides and give the same colour reactions (with Rhodamin G, dipikrylamine and cresyl violet). The same technique was used to determine semiquantitatively the amount of sulphatides in urinary sediments from patients with the actual disease as well as those with other neurological diseases, and from healthy infants, children and adults.

Austin (1957) concluded that a positive test for metachromatic lipids in urinary sediments (sulphatides) is pathognomonic for late infantile metachromatic leucodystrophy. On the other hand, we have found that sulphatides are always excreted in the urine of normal infants, children and adults. Healthy infants under 1 year excreted as much or even more than our patients with metachromatic leucodystrophy. However, these patients, aged 3-5 years, had much more sulphatides in their urine than normal children of the same age. We also found large amounts of urinary sulphatides in some children with probable vaccination encephalomyelo-

pathy, subacute progressive encephalitis, and some degenerative disorders of the central nervous system not hitherto verified histologically. Thus, diagnostic conclusions, drawn only from the excreted amounts of sulphatides, must be very cautious, especially as the significance of this test is further limited by the large variations in the daily excretion, making all quantitative estimations approximate. However, an additional alteration in the paper chromatogram can give a far better diagnostic support. Besides the large metachromatic sulphatide spot, a second more slow-running spot with the same colour characteristics was always found only in our patients with metachromatic leucodystrophy. This combined chromatographic pattern seems so far to be specific for the disease (Fig. 4).

Tests on Blood

Austin (1958) reported that metachromatic deposits also occur in white cells of the *peripheral blood* and bone-marrow of patients with metachromatic leucodystrophy. These abnormal cytoplasmic granules were found by Austin to be similar to those of Alder but distinct from ordinary 'toxic' granules. The granulocytic series is said to be more affected than lymphocytes or monocytes.

We have investigated the chemical composition of the *blood plasma*, with regard to the glycolipids, and found it to be of no diagnostic value. No significant differences from normal children were found either in the total amounts of the cerebroside or in the sulphatides (Hagberg and Svennerholm 1960). However, the cholesterol levels were slightly raised in the three cases examined.

Tests on Cerebrospinal Fluid

The cerebrospinal fluid has been found to be changed relatively early in the

disease, with a sometimes pronounced increase in the total protein without any simultaneous rise in the cell counts. Paper electrophoresis showed in our cases a normal relative distribution of the protein fractions. Except in diffuse cerebral sclerosis (type Krabbe), we have not seen this electrophoretic pattern in other demyelinating processes. Since metachromatic substances have also been found in damaged nerve roots (Hagberg *et al.* 1960), the increased protein content combined with a normal distribution of the electrophoretic components may possibly be ascribed to a nonspecific polyradicular irritation. In agreement with these findings is the clinical picture of the earlier stages, which is often very similar to the Guillain-Barré syndrome.

Sulphatides were present in the cerebrospinal fluid of our patients but the amounts were small and it was not possible to tell whether they were any greater than in normal specimens.

X-ray Findings

Cholecystography is of diagnostic interest, in this condition, for a silent non-functioning gall-bladder seems to be a characteristic finding, at least in the later stages (Brain and Greenfield 1950, Hagberg *et al.* 1960). A progressive loss of the gall-bladder's ability to concentrate could also be followed on repeated X-ray examination in one of our cases (Hagberg, Sourander and Svennerholm, *to be published*). At autopsy considerable fibrous thickening of the gall-bladder walls, cholesterosis and accumulation in the mucous folds of macrophages containing metachromatic and PAS-positive material has been found.

X-ray examination of the central nervous

system is of no diagnostic use in these cases. The only positive findings are the non-specific changes of cerebral atrophy in advanced cases.

Biopsy

Biopsies, taken mainly from three different locations, has been used to confirm the diagnosis during life.

Cortical biopsy was used in seven cases of metachromatic leucodystrophy by Blackwood and Cumings (1959). With chemical methods all seven were diagnosed while histological methods failed in three of them. None of the patients were any the worse for the operative procedure.

Biopsy of a peripheral nerve (N. musculocutaneus) with histochemical examination was found by Thieffrey and Lyon (1959) to be a useful aid for diagnosis in one case, verified by cortical biopsy. Our preliminary results from biopsies of the sural nerve point in the same direction (Fig. 5). Control material from cases with other demyelinating diseases is still lacking.

Renal biopsy was performed by Austin (1957), who verified one of his original cases in this way.

General Comments

In a fatal disease with no known treatment, early diagnosis is of doubtful value. But because this disease is genetically determined, an early correct diagnosis of the first affected child in a family is of great importance for future family planning. The disease is an 'inborn error of metabolism' and is probably due to a hitherto hypothetical dysfunction of an enzymatic process. In future it may be possible to detect the exact type of this defect both in patients and in carriers, and it may, perhaps, also be possible to correct it.

SUMMARY

Late infantile metachromatic leucodystrophy, a genetically determined neuropilidosis with accumulation of sulphatides in various organs, can now be diagnosed during life.

This paper surveys the clinical symptoms and signs, the laboratory tests in urine, blood and cerebrospinal fluid, the X-ray findings, and the diagnostic biopsies from brain cortex, peripheral nerves and kidney.

RÉSUMÉ

Diagnostic clinique et de laboratoire de la leucodystrophie métachromatique

Il est possible à présent de diagnostiquer, in vivo, la leucodystrophie métachromatique tardive du nourrisson, une neurolipidose déterminée génétiquement avec accumulation de sulfatides dans divers organes. On passe en revue: les symptômes et signes cliniques, les tests de laboratoire pratiqués sur l'urine, le sang et le liquide céphalorachidien, les résultats radiologiques, et les biopsies diagnostiques provenant du cortex cérébral, des nerfs périphériques et des reins.

ZUSAMMENFASSUNG

Klinische und laboratorische Diagnose der metachromatischen Leukodystrophie

Die spätinfantile metachromatische Leukodystrophie, eine genetisch bestimmte Neurolipidosis mit Anhäufung von Sulfatiden in verschiedenen Organen, kann jetzt bei Lebzeiten diagnostiziert werden. In diesem Artikel wird eine Übersicht über die klinischen Symptome und Zeichen, die Laborproben in Harn, Blut und Liquor, die Röntgenbefunde, und die diagnostischen Biopsien der Hirnrinde, der peripheren Nerven und der Nieren, gegeben.

REFERENCES

- Austin, J. H. (1957) 'Metachromatic form of diffuse cerebral sclerosis. I. Diagnosis during life by urine sediment examination.' *Neurology*, **7**, 415.
- (1957) 'Metachromatic form of diffuse cerebral sclerosis. II. Diagnosis during life by isolation of metachromatic lipids from urine.' *Ibid.*, **7**, 716.
- (1958) 'Observations in metachromatic leucoencephalopathy.' *Trans. Amer. neurol. Ass.*, 149.
- (1959) 'Metachromatic sulfides in cerebral white matter and kidney.' *Proc. Soc. exp. Biol. Med. (N. Y.)*, **100**, 361.
- (1960) 'Metachromatic form of diffuse cerebral sclerosis. III. Significance of sulfatide and other lipid abnormalities in white matter and kidney.' *Neurology*, **10**, 470.
- Blackwood, W., Cumings, J. N. (1959) 'Diagnostic cortical biopsy, a histological and chemical study.' *Lancet*, **ii**, 23.
- Brain, W. R., Greenfield, J. G. (1950) 'Late infantile metachromatic leucoencephalopathy, with primary degeneration of the interfascicular oligodendroglia.' *Brain*, **73**, 291.
- Greenfield, J. G. (1933) 'A form of progressive cerebral sclerosis in infants associated with primary degeneration of the interfascicular glia.' *J. Neurol. Psychopath.*, **13**, 289.
- Hagberg, B., Sourander, P., Svennerholm, L., Voss, H. (1959) 'Late infantile metachromatic leucodystrophy of genetic type.' *Acta paediat.*, **48**, 200.
- — — (1960) 'Late infantile metachromatic leucodystrophy of the genetic type.' *Ibid.*, **49**, 135.
- — — (1961) 'Sulfatide lipidosis in childhood.' (*To be published*)
- Svennerholm, L. (1959) 'Laboratory diagnostic tests in metachromatic leucodystrophy.' *Acta paediat.*, **48**, 632.
- (1960) 'Metachromatic leucodystrophy—a generalised lipidosis. Determination of sulfatides in urine, blood plasma and cerebrospinal fluid.' *Ibid.*, **49**, 690.
- Hain, R. F., LaVeck, G. D. (1958) 'Metachromatic leuco-encephalopathy. Review with illustrative case report.' *Pediatrics*, **22**, 1064.
- Hirsch, T. v., Peiffer, J. (1957) 'A histochemical study of the pre-lipid and metachromatic degenerative products in leucodystrophy.' In *Cerebral Lipidoses*, ed. Van Bogaert, Cumings and Lowenthal, Oxford: Blackwell, p. 68.
- Jatzkewitz, H. (1958) 'Zwei Typen von Cerebrosid-schwefelsäureestern als sog. "Prälipide" und Speichersubstanzen bei der Leukodystrophie, Typ Scholz (metachromatische Form der diffusen Sklerose).' *Hoppe-Seyler Z. physiol. Chem.*, **311**, 279.
- (1960) 'Die Leukodystrophie, Typ Scholz (metachromatische Form der diffusen Sklerose), als Sphingolipidose (Cerebrosid-schwefelsäureester-Speicher-Krankheit).' *Ibid.*, **318**, 265.
- Jefferson, M. (1958) 'Late infantile metachromatic leucodystrophy.' *Proc. R. Soc. Med.*, **51**, 160.
- Radin, N. S., Martin, F. B., Brown, J. R. (1957) 'Galactolipide metabolism.' *J. biol. Chem.*, **224**, 499.
- Thieffry, S., Lyon, G. (1959) 'Diagnostic d'un cas de leucodystrophie métachromatique (type Scholz) par la biopsie d'un nerf périphérique.' *Rev. neurol.*, **100**, 452.

Diagnosis of Metachromatic Leucodystrophy

After hearing Dr. Hagberg read the preceding paper at the N.S.S. Study Group in Oxford in 1960, Prof. J. N. Cumings wrote:

- (1) 'I have found on occasion that a positive result for metachromatic material is obtained in the urine of young infants, usually females, who are not sufferers from leucodystrophy. Clean or catheter specimens will probably be advisable.
- (2) Degrees of metachromasia depend on pH, and it may be that this feature has not been adequately controlled.
- (3) I have used the Cresyl Violet Acetic Acid staining technique of v. Hirsch and Pfeiffer as well as Toluidin Blue, and if anything prefer the former technique.'

These comments were shown to Dr. Hagberg and his colleagues, who reply:

- (1) 'In girls, catheter specimens will probably give more accurate results, but since repeated tests ought always to be done we do not like to do this. Specimens taken after thorough washing have been found satisfactory.
- (2) Concerning pH, Austin (*Neurology*, 1957, 7, 415) says: "Special pH precautions are not essential in the actual performance of fresh urine sediment examinations." We have not made any investigations of our own on this point. Personally I think that pH may play some role, but we have not found it necessary to complicate what is now an easily performed test, which we have always used in its original form as a screening test, combined with the much more accurate chemical examination for sulphatides. We never trust a single negative result of microscopy.
- (3) We have had no experience with Cresyl Violet instead of Toluidine Blue.'

NOTICE

Forward Trends

AN administrative curtain hangs between the 'educable' and the 'ineducable' child, as between the medical and educational professions. *Forward Trends in the Treatment of the Backward Child*, a quarterly published by the Guild of Teachers of Backward Children, seeks to bring together all the people working on behalf of backward children, irrespective of the kind or degree of their backwardness. It provides a forum in which the causes and possible educational treatment of backwardness can be discussed, pooling the experience of teachers, psychologists, doctors and social workers. Articles of up to 1,500 words are invited, and should be sent to the Editor, Mr. S. S. Segal, at 125 High Holborn, London, W.C.1. The annual subscription is 10s.

Acute Hemiplegia in Childhood

With a Note on the Management of the Acute Episode

MARTIN BAX

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THE National Spastics Society recently organised a study* group at Clevedon in Somerset. The subject was Acute Infantile Hemiplegia. This meeting was one of a series organised by the Society in which a relatively small group of people get together for informal discussions on various aspects of child neurology and cerebral palsy. The papers read at this conference, with an account of the discussions which arose, will shortly be published as a *Little Club Clinic in Developmental Medicine*. This article attempts to outline the main points made at the conference and to give a brief review of our present knowledge of acute hemiplegia in childhood.

Nomenclature

The nomenclature of acute hemiplegia in childhood has been confused in the past. Some writers have implied a distinction between acute hemiplegia of known aetiology and the syndrome of acute infantile hemiplegia, the aetiology of which was usually unknown. Cases of the latter syndrome sometimes fitted the classic conception of sudden onset accompanied by a convulsion, as described in current textbooks, but there were many exceptions. The use of the term 'acute infantile hemiplegia' is potentially dangerous as it tends to make the physician believe that in attaching

this label to any particular patient he has sufficiently exercised his diagnostic skill and the problem has been solved. It is preferable, therefore, to refer to the condition as acute hemiplegia and to attempt further elucidation of the prime cause.

Incidence

Hemiplegia, both acquired and so-called congenital, accounts for about a third of all cases of cerebral palsy, and of these hemiplegics about a quarter acquire their hemiplegia postnatally. It is still extremely difficult at times to say whether hemiplegia that is first recognised at the age of 6 months has been acquired postnatally or was acquired at or before birth.

Clinical Picture

The classic accounts of acute infantile hemiplegia describe the sudden appearance of hemiplegia at a variable time after a series of convulsions in a child who has previously been completely well or has only had a minor upset. The accompanying convulsions are sometimes limited to one side only, but are often generalised. The condition was graphically described by Freud (1897) and also by Osler (1889), who gave the following example.

Case 10. Rita O'N—, aet. 2 years and 4 months. H.285. When 15 months old, in April, during teething, had a convulsion; perfectly well before the attack, which came on suddenly and was confined to the left side and lasted 8 hours. In July she had a second attack, the convulsion lasting 20 hours. After the April attack there was complete paralysis of left arm, leg, and part of face; no strabismus. The second attack did not increase the paralysis. She had begun to recover power before the July convulsions. Reflexes increased on affected side.

* The participants in the Clevedon Study Group were: Martin Bax, Sven Brandt, A. Brodal, John Cavanagh, Philip Evans, John Marshall, Ian Mackenzie, Ross Mitchell, R. M. Norman, Tom Oppé, G. Pampiglione, P. E. Polani, Paul Sandifer, Peter Schurr, Kenneth Till.

Zofia Majewska sent a paper but did not attend.

In some cases, as Osler himself noticed, the paralysis comes on suddenly, without any preceding or associated general disturbance of health. Other cases follow acute infectious diseases such as measles, mumps or scarlet fever. In these, hemiplegia may develop at the onset of the acute illness or may appear some time after the onset (5 to 21 days). Most cases thus fall into one of three broad groups:

1. Onset accompanied by convulsions
2. Sudden onset without convulsions
3. Previous or present history of acute infectious disease.

Other cases of acute hemiplegia may be associated with conditions such as congenital malformations of the heart or tuberculous meningitis. In the past these have been considered separately from the syndrome of acute infantile hemiplegia, but, as has been mentioned, such a division is no longer considered valid.

Subsequent Clinical Course

Some cases of acute hemiplegia following convulsions will be instances of what is commonly known as 'Todd's paralysis,' a transient hemiplegia following epilepsy, which clears up usually within 24 hours, and nearly always within a week. At their onset, however, it is not possible to distinguish between these cases and those in which the hemiplegia is going to be permanent or, at least, longer lasting. Moreover, it is probably not desirable to make such a distinction, since present evidence suggests that the transient hemiplegias merely represent minor degrees of neuronal damage which, when more severe, may cause permanent hemiplegia.

When the hemiplegia has lasted more than a week the outlook is serious and there are likely to be residual defects in over three-quarters of the cases. The younger the patient the worse the outlook, and when in a child under 2 years there is very little likelihood of complete recovery. As a

rule the lower limb improves and the patient is able to walk; facial palsy also tends to improve but the outlook for the upper limb is worse and when this is involved there is almost always some residual defect. Possibly more serious than these motor defects is the distressingly high incidence of both mental defect and epilepsy in these children. About two-thirds of the patients develop epilepsy, and mental defect is very common, particularly where the epilepsy persists. In some series all the children over the age of 10 were mentally subnormal.

Aetiology

This is where our present ignorance is most apparent, for in very few cases it is possible to make a precise statement about the pathology of the condition. In children with congenital malformations of the heart, embolic phenomena are important. When such a child has a fit or develops hemiplegia, a cerebral abscess should always be excluded before a simple embolus is diagnosed; many of these abscesses are sterile by the time exploration is undertaken.

The cases associated with acute infectious disease presumably represent some type of allergic reaction but the exact nature of such reactions is unknown. The most likely cause for a sudden hemiplegia which develops spontaneously and is unaccompanied by a fit is a cerebral vascular accident; in some instances, angiography has demonstrated occlusion of a middle cerebral artery.

In the cases heralded by convulsions, our present knowledge derives largely from studies of the brains of patients who have died in status epilepticus. There is strong evidence, if not proof, that at least some of the neuronal destruction seen in these cases actually takes place during the convulsion. In long-standing epilepsy signs of old brain damage in the form of cortical loss and gliosis in the vulnerable areas are often associated with less conspicuous

signs of recent damage in the same areas. In other words, *brain damage may continue during life unless the convulsions are controlled.* In most fatal cases of status epilepticus the changes are bilateral, though one cerebral hemisphere may be more severely affected than the other; in a few fatal cases hemiplegia has been noted before death.

Investigations

When a child is admitted to hospital either in status epilepticus or with acute hemiplegia already established, investigations must be initiated without delay. The essential features of such investigations will be:—

1. A careful history.
2. A full general and neurological examination.
3. Careful observation, with regular recording of temperature, blood-pressure and respiration. (Variations in the blood-pressure may well be associated with, or relevant to, the development of permanent brain damage, and regular recordings should, therefore, be made. It is important always to record also the method and technique employed in measuring the blood-pressure.)
4. Acute infections should be recognised and, if possible, treated. Chest X-ray films and examination of the peripheral blood-picture may well be essential features of the routine.
5. The cerebrospinal fluid should always be examined, and a lumbar puncture should therefore be performed without delay.
6. Where the onset has been sudden and no aetiological factor can be identified, further investigations may have to be undertaken early in the course of the disease. This will often necessitate transfer to a specialist unit for angiography, air studies and EEG recording within a day or two of the onset.

Treatment

(1) The Convulsing Child

(Note. The hemiplegia may not be apparent or even present when the convulsions start. For this reason these suggestions regarding treatment must apply to all children admitted to hospital with convulsions of any kind.)

The convulsions must be stopped within half an hour of admission—McGreal (1957) suggests 15 minutes. Attempts to establish a diagnosis from the history and examination should, of course, be made soon after admission, but the pressing need is to control the seizure, and the following routine is suggested:

- (a) Paraldehyde should be injected intramuscularly at once (1.5 ml. paraldehyde per 10 lb. body-weight up to 6 ml.).
- (b) The air-passages should be cleared and a good airway maintained.
- (c) The stomach should be emptied.
- (d) If the paraldehyde is not effective after 30 minutes a general anaesthetic should be administered, preferably by an experienced anaesthetist; such is the seriousness of this condition, however, that if an anaesthetist is not available, administration should not be delayed. Inexperienced administrators should use the anaesthetic agent with which they are most familiar, and for many this will mean open ether; thiopentone is best avoided.
- (e) Arrangements should be made to give more paraldehyde intravenously, so as to maintain an anticonvulsant action after the anaesthetic has worn off. The paraldehyde drip in a young patient may necessitate a cut-down, and this can conveniently be carried out while the patient is still under the anaesthetic. (For intravenous use a 10 per cent solution of paraldehyde in normal saline is suggested; about 2 ml. in 20 ml. of saline is an average quantity

to use.) Intravenous paraldehyde should be continued as long as is necessary to prevent further convulsions.

- (f) Measures should be taken to control any hyperpyrexia and to maintain the blood-pressure and general condition.
- (g) Barbiturates should be avoided, at least as the first line of attack.
- (h) Antibiotics should be given if indicated. In most cases, the child can be observed for up to 24 hours after the hemiplegia develops, provided the fits have stopped. If the hemiplegia persists for longer, a neurologist's opinion should be sought, with a view to undertaking further investigations if these are considered advisable.

(2) *Acute Hemiplegia without Preceding Fits*

It is not always easy to be certain that the child has not had a convulsion—for example, when the parents give a history of finding the child hemiplegic when they went to get him up in the morning. However, if the initial investigations suggested above have been carried out and the aetiology is still in doubt there should be no delay in obtaining a neurologist's opinion on the need for special studies.

(3) *Hemiplegia associated with Acute Infectious Diseases*

These cases fall into two main categories:

- (a) There is a history of an exanthematous condition 5 to 21 days before the onset of hemiplegia. In this group a trial of steroids is recommended, but again, if the condition does not improve, neurological assistance should be sought.

- (b) Hemiplegia develops at the onset of the infection. As a rule these cases can safely be observed for 24 hours before further steps are taken.

Congenital Heart Disease

It is emphasised that the onset of hemiplegia or the occurrence of a fit in a child with a congenital malformation of the heart should be taken as evidence of the presence of a cerebral abscess until proved otherwise, and is no indication for initiating anticoagulant therapy.

Further Study

There are many gaps in our knowledge of acute hemiplegia in childhood. There are two main ways in which we can hope to widen our knowledge.

The first is by detailed and accurate observation and recording of every case that occurs, so that adequate statistical information about the conditions is available. (The Clevedon Group recommended getting up a working party to decide exactly what clinical records should be kept in each case.)

Secondly, too few of these cases have been adequately investigated in the past, and it is hoped that in future specialised investigations will be undertaken more often. The risks of most investigations are small compared with the seriousness of the sequelae of acute hemiplegia. More specialised methods of treatment may be given a clinical trial by research groups; these include the use of hypothermia, to reduce cerebral oedema, and fibrinolytic agents.

SUMMARY

This article briefly reviews our present knowledge of acute hemiplegia in children, which was recently the subject of a Study Group held at Clevedon, Somerset, for the National Spastics Society. An account of these discussions is to be published as a *Little Club Clinic in Developmental Medicine*.

An outline is here given of the nomenclature of acute infantile hemiplegia, its incidence, clinical picture and course, the aetiology, and the results of investigations. Detailed recommendations are made regarding the treatment of the acute episode.

RÉSUMÉ

Hémiplégie aiguë dans l'enfance

Cet article fait brièvement le point de nos connaissances actuelles sur l'hémiplégie aiguë chez l'enfant, cet état ayant été récemment le sujet d'une symposium tenu à Clevedon, Somerset, par le Groupement de Recherches Pédiatriques de la National Spastics Society.

On donne une vue d'ensemble de la nomenclature des incidences du tableau clinique et de l'évolution, de l'étiologie, des résultats de diverses investigations et des recommandations détaillées en ce qui concerne le traitement.

ZUSAMMENFASSUNG

Akute Hemiplegie im Kindesalter

Dieser Artikel bespricht kurz unsere hientigen Kenntnisse betreffs akuter Hemiplegie bei Kindern, ein Zustand, der vor kurzem das Thema einer in Clevedon, Somerset, von der Verbindung für Pädiatrische Forschungen der 'National Spastics Society' abgehaltenen Sitzung war.

Es wird ein Überblick über Nomenklatur, Inzidenz, klinisches Bild und Verlauf, Ätiologie und Resultate der Prüfungen gegeben und ausführliche Empfehlungen hinsichtlich der Therapie werden gemacht.

REFERENCES

- Freud, S. (1897) 'Die infantile Cerebrallähmung.' *In* Nothangel's *Specielle Pathologie und Therapie*, vol. 9 Vienna: Holder.
 McGreal, D. A. (1957) 'Convulsions in childhood.' M.D. Thesis, University of St Andrews.
 Osler, W. (1889) *The Cerebral Palsies of Childhood*. Philadelphia: Blakiston. pp. 26.

SHORT BIBLIOGRAPHY

- Bailey, O. T. (1960) 'The pathology of juvenile hemiplegia.' *GP (Kansas)*, 21, 88.
 Bonnal, J. (1959) 'Les indications neuro-chirurgicales dans l'hémiplégie cérébrale infantile.' *Concours med.*, 81, 3409.
 Christensen, E., Brandt, S. (1959) 'Recurrent acute infantile hemiplegia caused by a deep hemispheric angioma.' *Nord. Med.*, 62, 1574. (*In Danish*).
 Crothers, B., Paine, R. S. (1959) *The Natural History of Cerebral Palsy*. Cambridge, Mass.: Harvard University Press.
 Duffy, P. E. et al. (1957) 'Acute infantile hemiplegia secondary to spontaneous carotoid thrombosis.' *Neurology*, 7, 644.
 Ford, F. R. (1960) *Diseases of the Nervous System in Infancy, Childhood and Adolescence*. 4th ed. Oxford: Blackwell. pp. 882-6.
 — Schaffer, A. J. (1927) 'The etiology of infantile acquired hemiplegia.' *Arch. Neurol. Psychiat. (Chicago)*, 18, 323. (Reviews all the older literature and gives 154 refs.)
 Glander, R., Illert, H. (1958) 'Amaurose und Hemiparese nach Typhus abdominalis bei einem 3-jährigen Kinde.' *Arch. Kinderh.*, 158, 164.
 Grinberg, M., Neves, F. de P., Borges, M. A. (1959) 'Obstruction of the internal carotoid in a three-year-old child.' *Pediat. Prat. (S. Paulo)*, 30, 223. (*In Portuguese*).
 Hillcoat, B. L. (1959) 'Acute hemiplegia complicating cardiac disease in children.' *Med. J. Aust.*, ii, 604.
 Jirard, P., Aimard, Mme (1959) 'L'hémiplégie cérébrale infantile.' *J. Med. Lyon*, 40, 287.
 Mitchell, R. G. (1952) 'Venous thrombosis in acute infantile hemiplegia.' *Arch. Dis. Childh.*, 27, 95-104.
 Stevens, H. (1959) 'Carotoid artery occlusion in childhood.' *Pediatrics*, 23, 699.
 Stewart, R. M. (1948) 'Infantile cerebral hemiplegia—clinical features and pathological anatomy.' *Edin. med. J.*, 55, 488.
 Tichena, I. N., Evisikova, E. F., Makarov, V. N. (1958) 'Paroxysmal tachycardia with hemiplegia and infarction-like changes in the electrocardiogram in a 24-year-old child.' *Pediatrics, Moskva*, 41, 76-80. (*In Russian*).
 Wyllie, W. G. (1948) 'Acute infantile hemiplegia.' *Proc. ro. Soc. Med.*, 41, 459.

The Plantar Response in Infants and Children

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PROBABLY the best known physical sign in clinical neurology is that which has been associated with the name of Babinski since his original description in 1896. The complete Babinski, or extensor response, consists of extension of the proximal phalanx of the great toe, abduction or fanning of the other toes, and flexion at the knee and hip. This is the abnormal response in contrast with the normal or flexor response, which consists of flexion and adduction of all the toes of the foot.

This clinical sign is elicited by firm brisk stroking of the lateral aspect of the sole of the foot from the heel towards the toes with a sharp instrument such as a key, pencil, or thumb-nail. The well-established clinical fact that an effective stimulus must be painful was confirmed by Kugelberg (1948) with recordings of the afferent nerve impulses.

When the Babinski response is well established, sensory stimuli from various parts of the body may cause extension of the great toe. It is interesting to demonstrate the response on squeezing the ear, or pricking the chest. In some instances where such remote stimuli are effective the response may be observed in the contralateral foot. The ease with which upward movement of the great toe can be produced in certain cases led to the description of other ways of producing the response, and the originators' names were attached to these new 'signs'. Two of the best known 'signs' consist of hyperextension of the great toe on squeezing the calf (Gordon 1904), and on rubbing the shin (Oppenheim

1902). In Strümpell's reflex the response is produced by flexing the lower limb; Rossolimo elicited the response by upward tapping of the great toe; and the Mendel-Bechterew reflex is produced by tapping the cuboid bone in the foot. Wartenberg (1947) entertainingly described the development of these signs. They are only less certain ways of getting the response, and although in the days of anatomical cerebral localisation they were thought to have a place in the precise localisation of a lesion (Fulton 1943), this is now very doubtful.

There can be no doubt or confusion about a strongly positive extensor response, but the astute clinician needs to be able to detect those early changes of the response which indicate abnormality. That considerable experience and training are necessary to do this emphasises the fact that the plantar response is not just completely positive or negative. There is a gradation from the flexor to the extensor response, and in the beginning the first hint of abnormality is the observation that the movement of the great toe is not quite as definitely flexor on one side as on the other. The importance of detecting these early changes means that one must be careful to elicit the response just as described above, and must pay attention to the position of the limb, as the reflex is more easily obtained if the leg is extended at the knee and to the position of the head, for the sign may become more apparent when the head is turned away to the side opposite to that being tested (Walshe 1923).

The gradual change of a flexor to an extensor response so often seen in clinical practice suggests that the extensor response differs from the flexor response quantitatively rather than in any other way. The recent work of Landau and Clare (1959) supports this view. They concluded, on the basis of electromyographic studies, that the extensor response is a hyperactive flexor response produced by increased irradiation of the sensory stimulus provoking contraction of the extensor hallucis longus.

Pathology

The Babinski response was regarded for many years as a sign of a lesion of the pyramidal tract. There were early doubts about the anatomical connotations of the sign when it was observed to occur transiently in some patients in coma, and even to vary from flexor to extensor within seconds following the sequence of the apnoeic periods in patients with periodic respiration (observations quoted by Walsh 1957). Work by Nathan and Smith (1955) has confirmed the doubts. These writers correlated clinical signs with histological lesions in the central nervous system. They found that:

- (a) No relationship exists between the anatomical state of the cortico-spinal tracts and the form of the plantar response, and the plantar response may be found with histologically normal cortico-spinal tracts.
- (b) A lesion of one side of the spinal cord may be associated with a contralateral Babinski response.
- (c) Apparently identical lesions on the two sides of the spinal cord may be associated with a normal plantar response on one side and an abnormal response on the other.

The Babinski response does not, therefore, necessarily signify an anatomical lesion of the pyramidal tract. This conclu-

sion can be accepted, but not the inference that the plantar response is unreliable. Such inference assumes that a lesion of the pyramidal tract must be anatomically demonstrable and rejects the vast amount of experience from clinical practice which testifies to the value of the sign. Walshe (1956) strongly criticised the nihilistic approach of Nathan and Smith, and in his defence of the Babinski response also refuted the direct application of observations on animals to humans (Fulton and Keller 1932, Hoff and Beckenridge 1956). No attempt at summary can do justice to Walshe's article and the original text should be consulted.

This controversy about the Babinski response has been reviewed recently elsewhere (*Lancet* 1960), and has occurred during a period of realisation that the pyramidal system is a more extensive structure than it was considered to be previously (Lassek 1954). Thus, Lassek and others (1957) pointed out the discrepancy between the number of fibres in the corticospinal tract and the smaller number of Betz cells in the pre-Rolandic cortex. The appreciation of the ramifications of the pyramidal system fits in well with the current freer concept of cerebral physiology that is replacing earlier views.

Setting aside consideration of tracts and systems, the generalisation may be made that the Babinski response occurs as a result of the release of lower centres in the hind brain and spinal cord from cortical control. The finding of an extensor plantar response on one side indicates an interruption of the influence of the contralateral cerebral cortex. Its presence is no guide to the nature or to the extent of the lesion, which may be anatomically demonstrable, or functional, by which term is meant a disturbance, possibly biochemical, interfering with neuronal activity without producing permanent changes recognisable by our present histological techniques.

Clinical Significance

The Babinski sign is of considerable clinical value, as proved by long experience, and is particularly useful because false positive reactions are very seldom, if ever, encountered. The detection of an abnormal response always calls for further investigation to detect the cause.

The release of lower centres from cortical control that produces the extensor plantar response also produces spasticity so that the two are often found together. The Babinski response, however, should never be considered to be a sign of spasticity but more will be said about this later in connection with cerebral palsy in children.

The Babinski Response in Infancy

Attention has been devoted thus far to the concepts of adult neurology. These ideas have been applied successfully to older children, but for a long time it has been realised that the plantar response is almost invariably extensor in infancy. This is the basis of the teaching that the Babinski response is an unreliable sign under the age of two years. Over two the rules of adult neurology apply, but before then it doesn't matter! Paediatric medicine suffers from the application of the concepts of adult medicine to smaller individuals, and this is as true of neurology as of the other branches of medicine. In contrast to the above attitude the recent observations of Brain and Wilkinson (1959) show how rewarding can be the study of the Babinski response in the early years of life.

They carried out painstaking observations on 35 normal infants and 31 children with central nervous system lesions. Two types of stimuli, pin prick and cold, were applied to different parts of the body. They confirmed that an extensor response is found normally in infants. In neonates the response was produced by stimulation over a very wide area, but this receptive zone

decreased with age. Between 5 and 20 weeks of age stimulation of the sole of the foot and of the thigh regularly produced an extensor response, but stimulation of the trunk produced an extensor and flexor response with equal frequency. Not only did the receptive zone decrease as the babies got older but so also did the sensitivity to the cold stimulus. By 24 weeks of age cold had ceased to be an effective stimulus.

These observations by Brain and Wilkinson give to the 'physiological' extensor response of infancy a quantitative aspect, and show that it is useful not only to elicit the plantar response in the early years but to note the extent of the receptive zone. They concluded that their findings supported the theory of progressive centripetal myelination of the pyramidal tracts continuing during infancy. Thus it would seem reasonable to infer that the presence of a positive Babinski sign in a child with neurological disease from infancy might be due to persistence of the infantile plantar response with its characteristics of a wide receptive zone and response to a cold stimulus. It is disappointing that in the few children with cerebral palsy examined by Brain and Wilkinson (1959) the plantar responses obtained resembled more nearly those seen in adults with acquired spinal cord lesions than those of infants. This work needs repeating and extending. The measurement of the receptive zone and the response to different stimuli in certain groups of infants would be well worth while. What, for example, would be found in premature babies who are prone to develop spastic paraplegia. Woods (1957) discussed the hypothesis that this interesting condition might be due to incomplete myelination of the pyramidal tracts, but Polani (1958) suggested that the cause might be found in an incomplete maturation and migration of the nerve cells in the brain subserving motor function.

The Babinski Response in Cerebral Palsy

This is a suitable time to reconsider the status of the Babinski response in cerebral palsy. Examination for this sign is made in every patient with cerebral palsy, but it is not easy always to obtain a clear response, or to interpret the significance of the response observed. Although the concept that the infantile response gives place to the mature response during the second year of life can be accepted, it seems clinically that the response is not firmly established until later, so that in a young patient it may not be possible to be completely certain of the plantar response until they are a little older. This requires repeated examinations, but there is never anything lost by the reassessment of physical signs at intervals.

The feet of children with athetosis are very sensitive and stroking the foot usually intensifies the bizarre movements. Upward movement of the great toe may occur as part of the athetosis and be wrongly interpreted as a positive Babinski sign. But just as athetotic movements cause 'false positives' it must also be appreciated that an extensor plantar response can be seen in this type of cerebral palsy. Difficulty in obtaining the plantar response may also occur after surgical procedures on the feet, especially tendon transplantation.

Even when a definitely extensor plantar response is obtained in a child with cerebral palsy the significance of the sign has still to be assessed. In our present state of incomplete knowledge of this subject, it is better not to attempt to apply a preformed interpretation of the Babinski response: it is preferable to try to understand its meaning against the background of the other clinical features. This introduces a degree of vagueness that may be distasteful to those clinicians who wish to be very precise, but it leads towards a deeper and more satisfying appreciation of the neurological disturbances. Definite

statements may be made about some relationships of the plantar response, but others are admittedly speculative.

The presence of a Babinski response as a solitary physical sign does not indicate cerebral palsy. By definition cerebral palsy is a disorder of motor function, and the plantar response is not a test of motor function. However, the presence of an extensor plantar response in a child with a disorder of motor activity does indicate a central nervous system lesion, and thereby aids in the differential diagnosis between cerebral palsy and primary muscle disorders.

It follows from the above that the plantar response should always be considered with all the other physical signs. It is not a sign of primary importance in cerebral palsy but is secondary, or supplementary, to other signs which are the basis for the separation of the different clinical types. Thus, considering the two major categories of cerebral palsy, spasticity and athetosis, a diagnosis of the former is made on finding spastic hypertonus and an increased stretch reflex, and of the latter on observing abnormal movements. The majority of children with the spastic form of cerebral palsy do show an extensor plantar response, and the majority with athetosis show a flexor response. Although the knowledge of this relationship is a useful guide in clinical practice, the association is not invariable, and the Babinski response is not a sign of spasticity. Spastic cerebral palsy may be diagnosed in the absence of an extensor plantar response, and an extensor response may occasionally be found in other forms of cerebral palsy. This last fact was pointed out by Evans (1948), and has been re-emphasised by Crothers and Paine (1960) who wrote that 'the Babinski sign is in no way inconsistent with athetosis or pure extrapyramidal cerebral palsy'.

The Babinski sign is not related to the severity of the disease. It does not corres-

pond with the degree of functional disturbance or weakness nor, for that matter, is it any guide to prognosis.

The confusion that arises over the interpretation of the Babinski sign can be illustrated by reference to a child with infantile spastic hemiplegia who is found to have an upgoing toe on the 'normal' side, but no other evidence of involvement such as impaired movement, weakness, increased tendon reflexes, or clonus, on that side at all. Does the appearance of this sign on the 'normal' side indicate that that side is also affected, and should the child now be classified as triplegic or quadriplegic? I should have thought not, but those who retain the rigid 'upgoing toe = pyramidal tract lesion' philosophy of our student days might argue so. The arguments against this have been presented. The 'normal' side is normal in all respects apart from a single neurological sign and the child is still functionally hemiplegic.

It might be asked if this sign in the 'normal' leg indicates a bilateral cerebral lesion. This is not necessarily so but further evidence on this point might be obtained by using an observation by Brain and Wilkinson (1959) that a crossed extensor plantar response is more commonly associated with a bilateral cerebral lesion. It should be recalled that the crossed extensor plantar response is not just a Babinski response elicited by stimulating the opposite foot. The Babinski response when fully developed is accompanied by flexion of the knee and hip and is part of a flexor withdrawal reaction in response to a nociceptive stimulus, whereas the crossed extensor plantar response is associated with extension of the leg and is part of the crossed extension reflex.

An analogy might be drawn between the plantar response in this 'normal' leg and the abnormal electroencephalographic recording that may be obtained from the unaffected cortex in children with hemiplegia due to anatomically unilateral cerebral lesions. This abnormal rhythm often reverts to normal following hemispherectomy. If this operation were to be performed on the patient under discussion might the plantar response in the normal leg become normal?

Conclusion

There should be no fear that such a well-established physical sign as the Babinski response is losing status. The reconsideration and investigation of its role in children, particularly in those with cerebral palsy, will probably enhance its value especially when used in conjunction with other responses such as the crossed extension reaction. At the present time, in children over the age of two years, the Babinski response must be considered abnormal and indicative of central nervous system disturbances, serving thereby to distinguish between those with vague symptoms due to functional or emotional upsets and those with serious organic disease. The Babinski response needs to be interpreted with caution in cerebral palsy. In infancy the finding of an extensor response can be normal but the persistence of a wide receptive zone and response to cold may lead to the suspicion of a disturbance of the central nervous system. The sole of the foot is undoubtedly far from being the mundane object it may seem and will amply repay further attention.

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SUMMARY

The extensor plantar, or Babinski response consists of dorsiflexion of the great toe and fanning of the other toes, produced by firm painful stroking of the sole of the foot. This response is not always associated with an anatomical lesion of the pyramidal tract, as was

thought for a long time, but it is a very useful clinical sign indicating at least functional disturbance of the central nervous system. In all small babies the plantar response is extensor and can be elicited from a wide receptive zone. This zone diminishes as the infant grows older until towards the second year of age, when the normal response is flexor. The significance of the plantar response in cerebral palsy would bear careful re-examination.

RÉSUMÉ

La réponse plantaire dans l'infirmité motrice cérébrale

La réponse plantaire en extension, dite de Babinski, consiste en la flexion dorsale du gros orteil et en l'étalement des autres orteils par suite d'attouchements fermes et douloureux de la plante du pied. Cette réponse n'est pas toujours associée à une lésion anatomique du système pyramidal comme on le crut longtemps mais est une indication clinique très utile, tout au moins en ce qui concerne les troubles fonctionnels du système nerveux central. Chez tous les jeunes bébés, la réponse plantaire est en extension et peut être suscitée sur une zone réceptive étendue. Cette zone diminue au fur et à mesure que l'enfant grandit jusque vers deux ans, âge où la réponse normale est en flexion. Le sens à donner à la réponse plantaire, dans l'infirmité motrice cérébrale, aurait besoin d'être réexaminé avec soin.

ZUSAMMENFASSUNG

Die Fusssohlenantwort bei Zerebrallähmung

Die Fusssohlenantwort in Streckung oder Babinskisches Phänomen besteht aus Dorsiflexion der grossen Zehe und Spreizung der anderen Zehen nach festem und schmerz erregendem Bestreichen der Fusssohle. Diese Antwort ist nicht immer mit einer anatomischen Schädigung der Pyramidalbahn verbunden, wie man es lange geglaubt hat, ist aber ein sehr nützliches Klinisches Zeichen, wenigstens für die funktionellen Störungen des Zentralnervensystems. Bei allen Säuglingen ist die Fusssohlenantwort in Streckung und kann auf einem ausgedehnten empfänglichen Gebiete hervorgerufen werden. Dieses Gebiet nimmt, während das Kind wächst, nach und nach ab, bis um das zweite Lebensjahr, wo die normale Antwort Beugung wird. Es wäre nötig, die Bedeutung der Fusssohlenantwort bei Zerebrallähmung von Neuem eingehend zu studieren.

REFERENCES

- Babinski, M. J. (1896) 'Sur le réflexe cutané plantaire dans certaines affections organiques du système nerveux central.' *C.R. Soc. Biol. Paris*, 3, 207.
 Brain, R., Wilkinson, M. (1959) 'Observations on the extensor plantar reflex and its relationship to the functions of the pyramidal tract.' *Brain*, 82, 297.
 Crothers, B., Paine, R. S. (1959) *The Natural History of Cerebral Palsy*. London: Oxford University Press.
 Evans, P. R. (1948) 'Infantile cerebral palsy.' *Proc. roy. Soc. Med.*, 41, 402.
 Fulton, J. F. (1943) *Physiology of the Nervous System*, p. 412, 2nd ed. Oxford University Press.
 — Keller, A. D. (1932) *The Sign of Babinski, a Study of Cortical Dominance in Primates*. London: Baillière Tindall & Cox.
 Gordon, A. (1904) 'A new reflex: paradoxical flexor reflex; its diagnostic value.' *Amer. Med. Phila.*, 8, 97.
 Hoff, H. E., Beckenridge, C. G. (1956) 'The mammalian reflex prototype of the sign of Babinski.' *Brain*, 79, 155.
 Kugelberg, E. (1948) 'Demonstration of A and C fibre components in the Babinski plantar response and the pathological flexion reflex.' *Brain*, 71, 155.
Lancet (1960) 'The sign of Babinski.' I, 321.
 Landau, W. M., Clare, M. H. (1959) 'The plantar reflex in man, with special reference to some conditions, where the extensor response is unexpectedly absent.' *Brain*, 82, 321.
 Lassek, A. M. (1954) *The Pyramidal Tract: Its Status in Medicine*. American Lecture Series, No. 233. Springfield, Ill.
 — Woolsey, C. N., Walker, A. E., Boshes, B., Rose, A. S. (1957) 'Symposium of inquiry: The pyramidal tract.' *Neurology*, 7, 496.

- Nathan, P. W., Smith, M. C. (1955) 'The Babinski response; a review and new observations.' *J. Neurol., Neurosurg., Psychiat.*, **18**, 250.
- Oppenheim, H. (1902) 'Zur Pathologie der Hautreflexe an den unteren Extremitäten.' *Msehr. Psychiat. Neurol.*, **12**, 421; 518.
- Polani, P. E. (1958) 'Prematurity and "Cerebral Palsy".' *Brit. med. J.*, **ii**, 1497.
- Walsh, E. G. (1957) *Physiology of the Nervous System*. P. 375. London: Longmans Green.
- Walshe, F. M. R. (1914) 'The physiological significance of the reflex phenomena in spastic paralysis of the lower limbs.' *Brain*, **37**, 269.
- (1923) 'On variations in the form of reflex movements, notably the Babinski plantar response, under different degrees of spasticity and under the influence of Magnus and de Kleijn's tonic neck reflex.' *Brain*, **46**, 281.
- (1956) 'The Babinski plantar response, its form and its physiological and pathological significance.' *Brain*, **79**, 529.
- Wartenberg, R. (1947) 'The Babinski reflex after 50 years.' *J. Amer. med. Ass.*, **135**, 763.
- Woods, G. E. (1957) *Cerebral Palsy in Childhood*. P. 23. Bristol: Wright.

NOTICE

Memorial to William Little

THE London Hospital has lately had wall plaques erected in memory of two great men connected with the Hospital. One is on the house in Shoreditch where James Parkinson practised for 40 years and where he was living when he wrote his classic *Essay on the Shaking Palsy* in 1817. He had previously been a pupil of Richard Grindall, assistant surgeon to the London Hospital. The second plaque is on the wall of the Old Red Lion Inn,



in Leman Street, Aldgate, which stands on the site of an older Red Lion where William Little was born and where his father was the landlord. Little recalls an incident in his boyhood at the Inn, when he made friends with officers and men of the 10th Hussars billeted there on their way to the Battle of Waterloo: he must have been 5 at the time. The plaque to Dr. Little was unveiled by his grandson, Admiral Sir Charles Little, G.C.B. Five other grandsons were present at the ceremony.

Brain Function and the 5-Hydroxyindoles*

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IN 1954, Gaddum wrote: '... it is possible that the HT in our brains plays an essential part in keeping us sane...' His statement, based on evidence that I will outline, together with a similar pronouncement by Woolley and Shaw that same year, acted as a pistol shot to set going the vast avalanche of experimental work and speculation on hydroxytryptamine and allied compounds that has since swept into the world's journals.

When Page wrote a review on 'serotonin' (5-hydroxytryptamine, 5HT) in 1954 he was able to accumulate 153 references; when he brought the story up-to-date in 1958 he could refer to 529 papers.

Biologically, 5HT is widespread, large amounts being present in such diverse sites as the banana (Waalkes *et al.* 1958) and the male reproductive tract of the spiny dogfish (Mann 1960). In mammals, it is located in high concentration in the argentaffin cells of the gut—Bülbring and Lin (1958), in an elegant series of experiments, have shown that it may play an important part in the maintenance of normal gut motility. It is also well represented at certain sites in the central nervous system (Amin *et al.* 1954, Correale 1956, Bogdanski *et al.* 1957).

What, then, is the evidence that 5HT plays a part in the normal functioning of the central nervous system? Well, firstly it is *there*, in a distribution very similar to that of noradrenaline. It is largely concentrated in certain areas of grey matter—the hypothalamus, area postrema,

amygdala, caudate nucleus and midbrain—and this distribution is very similar in a number of different species that have now been investigated. Particularly high concentrations are present in the pineal gland (Giarman *et al.* 1960). Secondly, the enzyme which forms it, 5-hydroxytryptophan decarboxylase, is present in a broadly similar distribution.

Metabolism

Fig. 1 shows the 5-hydroxyindole pathway of tryptophan metabolism, for much of whose elucidation we are indebted to Udenfriend and his team at Bethesda (Udenfriend 1958). A variety of methods now exists for the assay of a number of these compounds in biological material (Sandler 1960). 5HT is produced from the amino-acid 5-hydroxytryptophan (5HTP) by the action of 5HTP decarboxylase. Although 5HTP has never been identified in the normal animal, it is sometimes excreted in high concentration in the urine of patients with a peculiar variety of carcinoid tumour, characterised by Sandler and Snow (1958). Patients with the more orthodox variety of tumour excrete large amounts of 5-hydroxyindoleacetic acid (5HIAA) only. 5HTP decarboxylase has been shown latterly to be the same enzyme as DOPA decarboxylase (Rosengren 1960a), important in the biosynthesis of the catechol amines. Although 5HT has been shown in tracer studies to derive from tryptophan (Udenfriend *et al.* 1956), the enzyme bringing about hydroxylation had not been identified until very recently, when Cooper and Melcer (1960) found it

* Based on a lecture given at the Fountain Hospital, London, on Dec. 8, 1960.

in extracts of rat and guineapig gut mucosa and demonstrated that the reaction would not occur unless ascorbic acid and copper were also present. This step is known to be the rate-limiting factor in 5HT production, for relatively enormous concentrations of 5HTP decarboxylase are present to bring about the reaction which follows it.

The action of another widespread enzyme, monoamine oxidase, present in high concentration in the hypothalamus, converts 5HT to 5-hydroxyindoleacetaldehyde, and this unstable intermediate is, in most species, straightway transformed to

5HIAA, the major excretory product. 5HT may also be eliminated as sulphate (Chadwick and Wilkinson 1960), glucuronide (Weissbach *et al.* 1958) or as an N-acetyl derivative (McIsaac and Page 1959), again depending on the species, although these are probably relatively minor pathways.

Sandler and others (1960) have recently demonstrated *in vitro* the existence of an alternative pathway by which 5HTP undergoes transamination, and the 5-hydroxyindolepyruvic acid thus formed is converted to 5HIAA (Fig. 1). This has since been confirmed by other workers

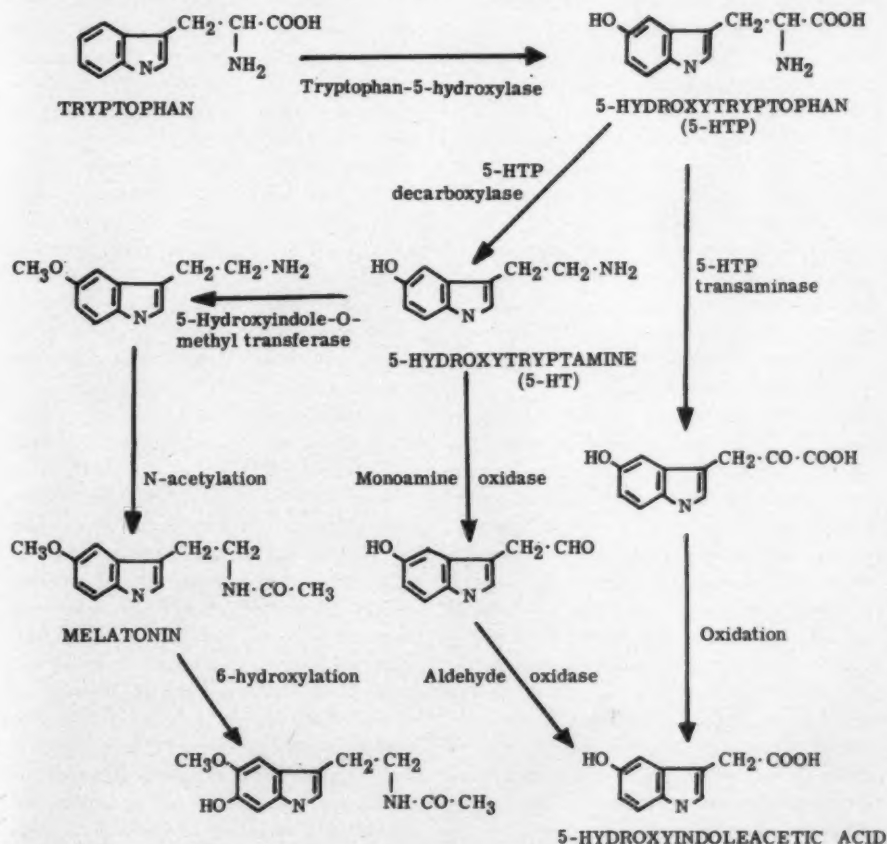


Fig. 1. The 5-hydroxyindole pathways of tryptophan metabolism.

(Spencer and Zamcheck 1960). We do not yet know how important this '5HT-shunt' mechanism is or whether it is active in vivo. So far we have only found it in rat liver and have not yet looked for it in brain. But 5HTP decarboxylase is so avid for 5HTP, and the turnover rate of 5HT is so great in brain—about 15 min. (Brodie *et al.* 1958)—that if present in brain the pathway is probably a minor one.

Another pathway of 5HT metabolism which has recently come into prominence is that via melatonin, N-acetyl-5-methoxytryptamine (Fig. 1; for review see Lerner and Case 1960). This compound, the most potent known lightening agent of frog skin, is found mainly in the pineal gland (Lerner *et al.* 1960). A specific methylating enzyme is found there (Axelrod and Weissbach 1960) together with a system producing N-acetylation (Weissbach *et al.* 1960). Its metabolic degradation is even more remarkable, occurring by hydroxylation at the 6-position on the indole ring (Kopin *et al.* 1960). Although traces of melatonin-like activity have been described in peripheral nerve tissue of man and other animals (Lerner *et al.* 1959), the function of melatonin in the nervous system remains obscure.

Although the turnover of 5HT in the brain is so great, its precise physiological role is similarly obscure. Gertner and others (1957) found traces of 5HT in the perfusate of the superior cervical ganglion of the cat after monoamine oxidase block with iproniazid, and this was associated with partial inhibition of transmission. Conversely, intracarotid 5HT can bring about cerebral synaptic inhibition (Gluckman *et al.* 1957).

How 5HT acts at cellular level is unknown, although Magnes and Hestrin-Lerner (1960) have recently indicated in perfusion experiments that it has an inhibitory effect on glycolysis. Woolley (1958) has suggested that it controls the

transport of calcium ions into the cells, and indeed, that all the biologically active amines exert their effects by regulating the passage of cations across cell membranes.

Clinical Studies

A great deal of our knowledge of central nervous system 5HT has stemmed from studies of the actions of drugs. A most important discovery in this field was that of Brodie and others (1955), who found that the alkaloid reserpine, which is used clinically for its sedative and hypotensive action, causes a marked and persistent fall in brain 5HT levels. Reserpine acts thus by blocking the binding of 5HT (Brodie *et al.* 1957), so that the free amine is rapidly metabolised by monoamine oxidase. It is not possible to explain all the pharmacological actions of reserpine in terms of 5HT depletion. For instance, its sedative action is not reversed by giving 5HTP (Carlsson *et al.* 1957), even though this substance is known to traverse the blood-brain barrier to be decarboxylated to 5HT in situ (Udenfriend *et al.* 1957). It is of interest that arousal *can* be achieved under these circumstances by giving dopa, which is decarboxylated to dopamine (Carlsson *et al.* 1957).

Analogues of reserpine, and allied drugs, have the power variably to deplete brain stores of 5HT and catechol amines (Finger *et al.* 1959). Although related compounds which have *no* sedative effect do not act as amine depleters (Brodie *et al.* 1956), it is still not easy to correlate quantitatively the clinical with the biochemical effects and to plot accurate structure-action relationships.

A group of drugs which provided a tremendous boost to research in this field is that typified by lysergic acid diethylamide (LSD). In minute concentration, this compound can produce temporary mental disturbance of a pattern which in some ways resembles schizophrenia (Stoll 1947).

When Gaddum (1953) found that LSD was an extremely efficient blocking agent to the action of 5HT the possibility arose that an endogenous LSD-like substance might be the cause of schizophrenia.

Kety (1959), in a critical review of current theories of schizophrenia, marshalled sufficient evidence to prove most of them untenable. However, he could not dismiss the possible significance of 5HT in the aetiology of this disease, even though concrete evidence to support its role has been tardy in arriving. It has even been difficult, for instance, to prove that LSD induces a model psychosis by antagonising 5HT—other compounds, such as brom-LSD, which antagonise 5HT just as well in vitro (Cerletti and Doepfner 1958), do not have any hallucinogenic action.

My colleagues and I (Bregelmann *et al.* 1958) did find, however, that pretreatment of normal volunteers with 5HTP tended to attenuate some of the effects of LSD, when measured by objective psychometric tests. But injection of 5HTP failed to increase performance scores in a group of schizophrenics who were put through similar tests (Bregelmann *et al.* 1959). Other workers have attempted to treat schizophrenic patients with 5HTP in fairly high dosage, with a similar lack of success (Hoagland 1958). It is not surprising. Schizophrenia is a name for a collection of symptoms which are probably merely the end-result of a diversity of noxious stimuli. We cannot, however, dismiss a trial of 5HTP completely, for there is always the possibility that within this heterogeneous collection there is a small group in which an endogenous LSD-like substance exists whose avidity for receptors is not too great to be displaced by excess 5HT.

It is of interest that 5HTP itself, when given to an experimental animal in very high dosage, can produce many of the signs that have been associated with LSD intoxication (Bogdanski *et al.* 1958).

This effect is due to large amounts of 5HT being formed in the brain; it can be brought about with much smaller 5HTP dosage if the monoamine oxidase mechanism which inactivates 5HT is previously inhibited.

Another link between the psychoses and 5HT is the ability of some monoamine oxidase inhibiting drugs to alleviate depression. Pare and I (1959) carried out a controlled clinical trial of one of these drugs, iproniazid, in a group of depressives and found 12 out of 50 whose improvement was due to the drug—i.e., who relapsed when treatment was withdrawn and improved when it was resumed. A characteristic feature in the response to the monoamine oxidase inhibitors is a lag of 5–7 days before clinical improvement is observed. It is therefore difficult to ascribe this improvement to a direct rise in brain 5HT; for brain 5HT levels increase in a matter of hours after the drug has been given. To test this point further, however, we treated patients who were known reactors to iproniazid with both 5HTP and dopa parenterally during the lag period but failed to obtain any shortening of it. An explanation may lie in the fact that other enzyme systems are affected as well as monoamine oxidase. It is likely too that other amines are metabolised in the brain by monoamine oxidase, so that the replacement of 5HT and dopamine alone was insufficient to restore the biochemical *status quo*.

There are still more data to associate mental disease with a disturbance in indole metabolism. My colleagues and I have demonstrated a disturbance of indole metabolism in certain groups of mental defectives which can broadly be classified as phenylketonurics and 'the rest'.

Although the fundamental defect in phenylketonuria—an inborn error of metabolism transmitted by a single recessive gene—is known to be an inability to

hydroxylate phenylalanine to tyrosine, there is no obvious explanation for the mental defect. (For review, see Crome and Pare 1960). Certainly phenylalanine itself is not directly toxic, although many abnormal metabolites which occur in the disease have been claimed to be toxic on rather exiguous evidence. It therefore occurred to us (Pare *et al.* 1957) that an inability to hydroxylate tryptophan might also be present. Whether this hypothesis is the true explanation or not, a marked decrease in circulating 5HT and urinary 5HIAA was observed in phenylketonurics when compared with controls and there was very little overlap in values (Pare *et al.* 1959a).

We found that these low values revert to normal when the patient is placed on a low phenylalanine diet (Pare *et al.* 1958), the only treatment so far available which gives any hope of normal development if it is started early enough in life.

We had been able to demonstrate by this time that 5HTP decarboxylase was inhibited *in vitro* by certain aromatic acid metabolites of phenylalanine which are excreted in abnormally large amounts in phenylketonuria (Davison and Sandler 1958), and we therefore devised an *in-vivo* test of decarboxylase activity, estimating urinary 5HT and 5HIAA after intravenous injection of 5HTP (Pare *et al.* 1958). Although the decreased output of these compounds in phenylketonurics suggested an impairment of their decarboxylase mechanism when compared with findings in matched controls, the investigation must be repeated, in the light of some recent data, in conditions of maximal urine acidity; for Sandler and Spector (*to be published*) have recently shown that the excretion of 5HT by a direct renal decarboxylation mechanism such as this is dependent on the pH of the excreted urine.

Whatever the mechanism of the deficiency of 5HT in phenylketonurics, it is unlikely

to be the sole cause of the mental defect. We carried out a small pilot trial of 5HTP in eight young phenylketonurics but were unable to record any improvement in their mental state after a period of 6 months (Pare *et al.* 1959b).

When we did our original work on the phenylketonurics, we were surprised to find that some of the serum 5HT levels in non-phenylketonuric children from the same hospital showed very high values. Sometimes the levels obtained were as high as those found in patients with the carcinoid syndrome.

My colleagues and I (Pare *et al.* 1960) have analysed a series of 83 non-phenylketonuric mental defectives from various hospitals and outpatient departments. Fig. 2 shows mean and standard error of the mean serum levels of 5HT for different disease entities, so far as they could be decided. Values represented by unshaded columns do not differ significantly from normal values. Among these is the chromosomal disease, mongolism. It is of interest that the only patient in this group with a high serum 5HT level had suffered a severe anoxic incident at birth. Data shown by stippled columns differed significantly from normal at the 1 in 1,000 level; stippled columns at the 1 in 20 level. These groups contained patients with mental defect of both genetic and environmental origin. We could find no correlation between 5HT level and degree of physical handicap. The only thing that these patients had in common was an I.Q. of less than 50. A group of patients with cerebral palsy and normal I.Q. was matched with a low I.Q. group with a similar degree of physical handicap. The patients with the normal I.Q. had normal serum-levels of 5HT. An interesting observation was that a group with a secondary dementia due to anoxia also tended to have a high serum 5HT.

There was no particular drug ingestion

pattern in any of these groups, and conversely, a series of epileptics of normal I.Q. who were taking large doses of phenytoin, which has been claimed to raise brain 5HT levels (Bonnycastle *et al.* 1957), had normal serum-levels of 5HT.

Although our group included mentally defective children living at home, it is just possible that the rise in 5HT may stem from some unaccounted for dietary factor; for Todrick and his colleagues (1960) have recently described high 5HT levels in some hospitalised psychotics.

The abnormal 5HT pattern was broadly reflected by 5HIAA excretion, so that the

most likely explanation for the increased serum 5HT levels is an increase in its production. Although there was a significant rise in platelet-count in such patients, the platelets which were present also had a higher saturation of 5HT.

Whether the increased 5HT production derives from the injured brain direct or is due to some release of gut argentaffin cells from brain inhibition must be decided in the future. Meanwhile, assay of serum 5HT level may be of some value in helping to confirm a diagnosis of mental defect in a child with cerebral palsy, often a very difficult clinical decision to make.

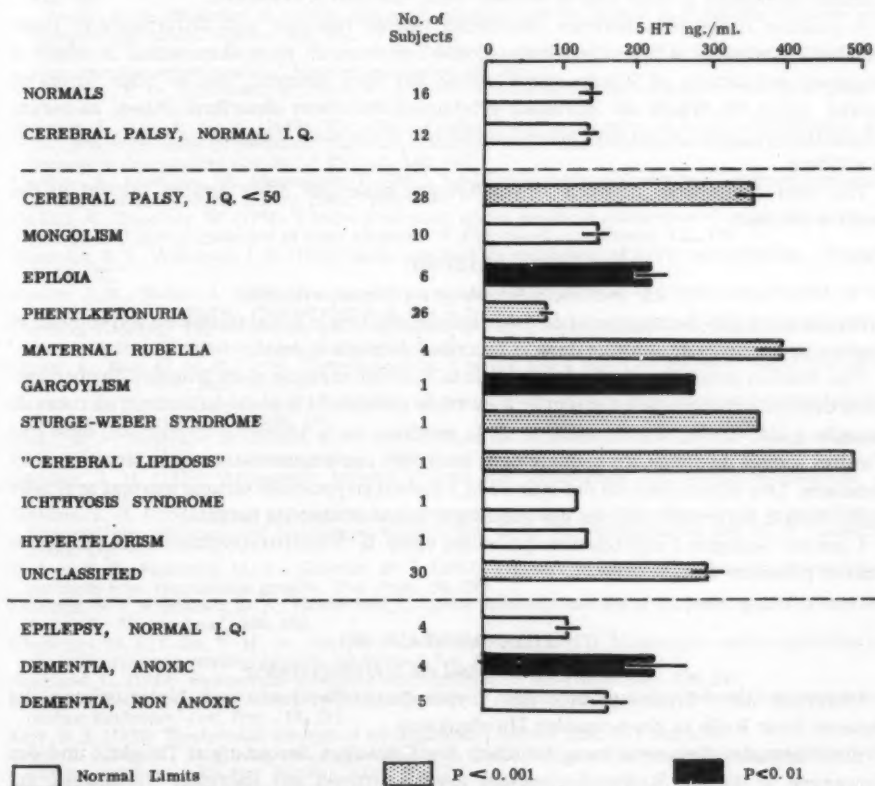


Fig. 2. Mean and standard error of the mean serum 5-hydroxytryptamine (5HT) levels in various groups of subjects with normal intelligence and with mental handicap.

Conclusions

What conclusions can be drawn from the large body of information I have tried to summarise? We seem to be no nearer to answering the question, 'What is the role of 5HT in normal brain function?'. I would suggest that the main lesson to be learnt from the present chaotic situation is that we have been focusing our attention too hard on two amines—5HT and noradrenaline—and have got the problem out of perspective. We must gather similar

information about dopamine (Rosengren 1960b), tryptamine (Sjoerdsma *et al.* 1959), the three tyramines (Sjoerdsma *et al.* 1960), octopamine (Kakimoto and Armstrong 1960), histamine (White 1960) and other amines which have been detected in urine but whose presence in brain is only just beginning to be suspected.

It is likely that the solution to the problem of the biochemical basis of the psychoses and possibly of mental defect lies somewhere in this direction.

SUMMARY

A review of the 5-hydroxyindole pathways of tryptophan metabolism is presented and evidence pertaining to their rôle in normal brain function is examined.

A possible connection between disordered mental function and disturbance in these metabolic pathways is discussed, with special reference to phenylketonuria, in which a decreased production of 5-hydroxytryptamine has been detected, and to other forms of mental defect, in which an increased production has been described. Assay of serum 5-hydroxytryptamine levels may prove a useful test in the early diagnosis of mental handicap in children.

The inter-relationship between 5-hydroxytryptamine and other amines present in the brain is stressed.

RÉSUMÉ

La fonction cérébrale et les 5-hydroxyindoles

Présentation des cheminements du 5-hydroxyindole dans le métabolisme du tryptophane, et examen des preuves de son rôle dans la fonction cérébrale normale.

Une relation possible entre les troubles de la fonction mentale et les troubles du cheminement dans ce métabolisme est discutée, avec étude spéciale de la phénylcétonurie, au cours de laquelle a été décelée une diminution de la sécrétion de la 5-hydroxytryptamine, ainsi que d'autres formes de déficience mentale dans lesquelles une augmentation de la sécrétion était constatée. Des déterminations des taux de la 5-hydroxytryptamine sérique peuvent se révéler utiles dans le diagnostic précoce des handicaps mentaux chez les enfants.

L'auteur souligne l'interrelation qui existe entre la 5-hydroxytryptamine et les autres amines présentes dans le cerveau.

ZUSAMMENFASSUNG

Die Hirnfunktion und die 5-Hydroxyindole

Übersicht der 5-Hydroxyindole des Tryptophanstoffwechsels und Untersuchung der Beweise ihrer Rolle in der normalen Hirnfunktion.

Ein eventueller Zusammenhang zwischen den Störungen der geistigen Tätigkeit und den Störungen in diesem Stoffwechselwandel werden erörtert mit spezieller Verweisung auf Phenylketonuria, bei der eine Verminderung der 5-Hydroxytryptaminerzeugung entdeckt wurde, sowie auf andere Schwachsinnformen, bei denen eine Zunahme der Sekretion

beschrieben wurde. Prüfungen des 5-Hydroxytryptamingehalts im Serum können sich für frühzeitige Diagnose der geistigen Defekte bei Kindern nützlich erweisen.

Die gegenseitigen Beziehungen zwischen 5-Hydroxytryptamin und anderen im Gehirn vorhandenen Aminen werden betont.

REFERENCES

- Amin, A. H., Crawford, T. B. B., Gaddum, J. H. (1954) 'The distribution of substance P and 5-hydroxytryptamine in the central nervous system of the dog.' *J. Physiol.*, **126**, 596.
- Axelrod, J., Weissbach, H. (1960) 'Enzymatic o-methylation of N-acetylserotonin to melatonin.' *Science*, **131**, 1312.
- Bogdanski, D. F., Weissbach, H., Udenfriend, S. (1957) 'The distribution of serotonin, 5-hydroxytryptophan decarboxylase, and monoamine oxidase in brain.' *J. Neurochem.*, **1**, 272.
- Bogdanski, D. F., Weissbach, H., Udenfriend, S. (1958) 'Pharmacological studies with the serotonin precursor, 5-hydroxytryptophan.' *J. Pharmacol. exp. Therap.*, **122**, 182.
- Bonnycastle, D. D., Giarman, N. J., Paasonen, M. K. (1957) 'Anticonvulsant compounds and 5-hydroxytryptamine in rat brain.' *Brit. J. Pharmacol.*, **12**, 228.
- Brengelmann, J. C., Pare, C. M. B., Sandler, M. (1958) 'Alleviation of the psychological effects of LSD in man by 5-hydroxytryptophan.' *J. ment. Sci.*, **104**, 1237.
- — — (1959) 'Effects of 5-hydroxytryptophan on schizophrenia.' *J. ment. Sci.*, **105**, 770.
- Brodie, B. B., Pietscher, A., Shore, P. A. (1955) 'Evidence that serotonin has role in brain function.' *Science*, **122**, 968.
- Shore, P. A., Pietscher, A. (1956) 'Serotonin releasing activity limited to Rauwolfia alkaloids with tranquillizing action.' *Science*, **123**, 992.
- Spector, S., Kuntzman, R. G., Shore, P. A. (1958) 'Rapid biosynthesis of brain serotonin before and after reserpine administration.' *Naturwissenschaften*, **45**, 243.
- Tornich, E. G., Kuntzman, R., Shore, P. A. (1957) 'On the mechanism of action of reserpine: effect of reserpine on capacity of tissues to bind serotonin.' *J. Pharmacol. exp. Therap.*, **119**, 461.
- Bülbring, E., Lin, R. C. Y. (1958) 'The effect of intraluminal application of 5-hydroxytryptamine and 5-hydroxytryptophan on peristalsis; the local production of 5HT and its release in relation to intraluminal pressure and propulsive activity.' *J. Physiol.*, **140**, 381.
- Carlsson, A., Lindqvist, M., Magnusson, T. (1957) '3,4-Dihydroxyphenylalanine and 5-hydroxytryptophan as reserpine antagonists.' *Nature, Lond.*, **180**, 1200.
- Cerletti, A., Doepfner, W. (1958) 'Comparative study on the serotonin antagonism of amide derivatives of lysergic acid diethylamide and of ergot alkaloids.' *J. Pharmacol. exp. Therap.*, **122**, 124.
- Chadwick, B. T., Wilkinson, J. H. (1960) 'Some aspects of the metabolism of 5-hydroxytryptamine.' *Biochem. J.*, **76**, 102.
- Cooper, J. R., Melcer, I. (1960) 'The enzymatic oxidation of tryptophan to 5-hydroxytryptophan in the biosynthesis of serotonin.' *Pharmacologist*, **2**, No. 2, p. 67.
- Correale, P. (1956) 'The occurrence and distribution of 5-hydroxytryptamine (enteramine) in the central nervous system of vertebrates.' *J. Neurochem.*, **1**, 22.
- Crome, L., Pare, C. M. B. (1960) 'Phenylketonuria: a review and a report on the pathological findings in four cases.' *J. ment. Sci.*, **106**, 862.
- Davison, A. N., Sandler, M. (1958) 'Inhibition of 5-hydroxytryptophan decarboxylase by phenylalanine metabolites.' *Nature, Lond.*, **181**, 186.
- Finger, K. F., Hughes, F. B., Brodie, B. B. (1959) 'Differential effects of various reserpine analogs on amines in central and peripheral nervous systems.' *Fed. Proc.*, **18**, 388.
- Gaddum, J. H. (1953) 'Antagonism between lysergic acid diethylamide and 5-hydroxytryptamine.' *J. Physiol.*, **121**, 15P.
- Gaddum, J. H. (1954) 'Drugs antagonistic to 5-hydroxytryptamine.' In Ciba Foundation Symposium on Hypertension: Humoral and Neurogenic Factors. Ed. G. E. W. Wolstenholme and M. P. Cameron. London. p. 75.
- Gertner, S. B., Paasonen, M. K., Giarman, N. J. (1957) 'Presence of 5-hydroxytryptamine (serotonin) in perfusate from sympathetic ganglia.' *Fed. Proc.*, **16**, 299.
- Giarman, N. J., Freedman, D. X., Picard-Ami, L. (1960) 'Serotonin content of the pineal glands of man and monkeys.' *Nature, Lond.*, **186**, 480.
- Gluckman, M. I., Cohn, V. H., Jr., Hart, E. R., Marrazzi, A. S. (1957) 'Monoamine oxidase inhibition (in situ serotonin) and cerebral synaptic inhibition.' *Fed. Proc.*, **16**, 47.
- Hoagland, H. (1958) 'Biochemical aspects of schizophrenia.' *J. nerv. ment. Dis.*, **126**, 211.
- Kakinoto, Y., Armstrong, M. D. (1960) 'Identification of octopamine in animals treated with monoamine oxidase inhibitors.' *Fed. Proc.*, **19**, 295.
- Kety, S. S. (1959) 'Biochemical theories of schizophrenia.' *Science*, **129**, 1528 and 1590.
- Kopin, I. J., Pare, C. M. B., Axelrod, J., Weissbach, H. (1960) '6-Hydroxylation, the major metabolic pathway for melatonin.' *Biochim. Biophys. Acta.*, **40**, 377.
- Lerner, A. B., Case, J. D. (1960) 'Melatonin.' *Fed. Proc.*, **19**, 590.
- Case, J. D., Mori, W., Wright, M. R. (1959) 'Melatonin in peripheral nerve.' *Nature, Lond.*, **183**, 1821.
- Takahashi, Y. (1960) 'Isolation of melatonin and 5-methoxyindole-3-acetic acid from bovine pineal glands.' *J. biol. Chem.*, **235**, 1992.

- McIsaac, W. M., Page, I. H. (1959) 'The metabolism of serotonin (5-hydroxytryptamine).' *J. biol. Chem.*, **234**, 858.
- Magnes, J., Hestrin-Lerner, S. (1960) 'Effect of 5-hydroxytryptophan and serotonin on glucose content and functional activity of the perfused cat brain.' *J. Neurochem.*, **5**, 128.
- Mann, T. (1960) 'Serotonin (5-hydroxytryptamine) in the male reproductive tract of the spiny dogfish.' *Nature, Lond.*, **188**, 941.
- Page, I. H. (1954) 'Serotonin (5-hydroxytryptamine).' *Physiol. Rev.*, **34**, 563.
- (1958) 'Serotonin (5-hydroxytryptamine); the last four years.' *Physiol. Rev.*, **38**, 277.
- Pare, C. M. B., Sandler, M. (1959) 'A clinical and biochemical study of a trial of iproniazid in the treatment of depression.' *J. Neurol. Neurosurg. Psychiat.*, **22**, 247.
- Stacey, R. S. (1957) '5-hydroxytryptamine deficiency in phenylketonuria.' *Lancet*, **i**, 551.
- — (1958) 'Decreased 5-hydroxytryptophan decarboxylase activity in phenylketonuria.' *Lancet*, **ii**, 1099.
- — (1959a) 'The relationship between decreased 5-hydroxyindole metabolism and mental defect in phenylketonuria.' *Arch. Dis. Child.*, **34**, 422.
- — (1959b) '5-hydroxytryptamine deficiency in phenylketonuria; its relation to the abnormalities of phenylalanine metabolism and to the associated mental defect.' In *Neuro-Psychopharmacology*. ed. P. B. Bradley, P. Beniker and C. Radouco-Thomas. Elsevier, Amsterdam. p. 648.
- — (1960) '5-Hydroxyindoles in mental deficiency.' *J. Neurol. Neurosurg. Psychiat.*, **23**, 341.
- Rosengren, E. (1960a) 'Are dihydroxyphenylalanine decarboxylase and 5-hydroxytryptophan decarboxylase individual enzymes.' *Acta. physiol. scand.*, **49**, 364.
- (1960b) 'On the role of monoamine oxidase for the inactivation of dopamine in brain.' *Acta. physiol. scand.*, **49**, 370.
- Sandler, M. (1960) 'Laboratory investigation of abnormal indole metabolism.' Chapter 12 in *Recent Advances in Clinical Pathology*. Ed. S. C. Dyke. Series III. London: Churchill, p. 163.
- Snow, P. J. D. (1958) 'An atypical carcinoid tumour secreting 5-hydroxytryptophan.' *Lancet*, **i**, 137.
- Spector, R. G., Ruthven, C. R. J., Davison, A. N. (1960) 'The occurrence and adaptive increase of 5-hydroxytryptophan- α -oxoglutarate transaminase in rat liver.' *Biochem. J.*, **74**, 42P.
- Sjoerdsma, A., Oates, J. A., Gillespie, L., Jr. (1960) 'Quantitation of monoamine oxidase inhibition produced with various drugs in man.' *Proc. Soc. exp. Biol., N.Y.*, **103**, 485.
- Zaltsman, P., Udenfriend, S. (1959) 'Identification and assay of urinary tryptamine: application as an index of monoamine oxidase inhibition in man.' *J. Pharmacol. exp. Therap.*, **126**, 217.
- Spencer, R. P., Zamcheck, N. (1960) 'Alternate metabolic fate of 5-hydroxytryptophan.' *Biochem. Biophys. Res. Comm.*, **3**, 386.
- Stoll, W. A. (1947) 'Lysergsaure-diäthylamid, ein Phantastikum aus der Mutterkorngruppe.' *Schweiz. Arch. Neurol. Psychiat.*, **60**, 1.
- Todrick, A., Tait, A. C., Marshall, E. F. (1960) 'Blood platelet 5-hydroxytryptamine levels in psychiatric patients.' *J. ment. Sci.*, **106**, 884.
- Udenfriend, S. (1958) 'Metabolism of 5-hydroxytryptamine.' In *5-Hydroxytryptamine*. Ed. G. P. Lewis. London: Pergamon Press, p. 43.
- Titus, E., Weissbach, H., Peterson, R. E. (1956) 'Biogenesis and metabolism of 5-hydroxyindole compounds.' *J. biol. Chem.*, **219**, 335.
- Weissbach, H., Bogdanski, D. F. (1957) 'Increase in tissue serotonin following administration of its precursor 5-hydroxytryptophan.' *J. biol. Chem.*, **224**, 803.
- Waalkes, T. P., Sjoerdsma, A., Creveling, C. R., Weissbach, H., Udenfriend, S. (1958) 'Serotonin, norepinephrine and related compounds in bananas.' *Science*, **127**, 648.
- Weissbach, H., Redfield, B. G., Axelrod, J. (1960) 'Biosynthesis of melatonin: enzymic conversion of serotonin to N-acetylserotonin.' *Biochim. Biophys. Acta*, **43**, 352.
- Udenfriend, S. (1958) 'Serotonin -o-glucuronide; an alternative route of serotonin metabolism.' *Fed. Proc.*, **17**, 418.
- White, T. (1960) 'Formation and catabolism of histamine in cat brain in vivo.' *J. Physiol.*, **152**, 299.
- Woolley, D. W. (1958) 'A probable mechanism of action of serotonin.' *Proc. nat. Acad. Sci., Wash.*, **44**, 197.
- Shaw, E. (1954) 'A biochemical and pharmacological suggestion about certain mental disorders.' *Ibid*, **40**, 228.

The Study of Imbecile Behaviour

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A SOCIETY's concern for the handicapped is related not only to humanitarian trends within it, but also to prevailing economic conditions which largely determine the degree of social welfare and security afforded to its members. The last war gave a great impetus to the rehabilitation of the physically handicapped, while the post-war years of full employment have seen an increasing interest in the problems of the mentally subnormal and disordered. As a result, the mildly subnormal are increasingly dealt with in the community, and hostels and rehabilitation centres are gradually becoming available for their care and training.

The severely subnormal,* however, present a different problem, since by their nature they are permanently incapable of leading a truly independent existence. Furthermore, there is impressive evidence to suggest that their numbers are increasing, as a result of medical and social advances which have led to a decline in infant mortality and prolonged life span. Benda (1952) showed that in New York State 19 children per thousand who would have died from 'congenital debility and malformation' in 1920, survived in 1949. Obviously some of these would be

imbeciles and idiots. More recently Carter (1958) has shown that in 1929, one in every 4,000 ten-year-old children was a mongol; in 1949 the proportion had doubled and one in every 2,000 ten-year-olds was a mongol, while today it has doubled again and one in every thousand ten-year-olds is thus affected. Since the birth-rate of mongolism is about one in every 600 live births, it seems probable that the present trend, if it continues, will lead to an even greater number of mongols reaching this age, and probably beyond it. The problem of alleviating the burden of severe handicap in the family and in society is thus becoming urgent.

During the last century an increasing interest has been accorded to the medical and social aspects of a variety of physical and mental disabilities such as blindness, deafness, cerebral palsy, and mental disorder and subnormality. Two main trends characterise attempts to alleviate such conditions: on the one hand, research into causes and the use of surgical and chemotherapeutic techniques; on the other hand, the development and use of remedial training, which although not 'curing' the condition, often serves substantially to control or modify its manifestations. While complete prevention or cure is the ultimate hope, and while the requisite knowledge is being acquired, importance is attached to educational techniques directed towards the best adjustment of the handicapped individual in society. This usually demands the thorough understanding of the behavioural potentials of the indi-

* The Mental Health Act (1959) brings into use the term 'severely subnormal', which includes all those formerly classified as idiots, imbeciles and some of the lower grades of feeble-mindedness. It is unfortunate that the considerable differences in ability existing between such grades of subnormality are to some extent obscured by the use of a 'blanket' term. In this article, therefore, severe subnormality is used synonymously with imbecility, which is its major constituent.

vidual and the development of new skills and the exploitation of assets which might not normally assume great importance and indeed might not normally be visible.

So far as imbeciles are concerned, attention has been focused on three main aspects (apart from the genetic and the medical): the problem of the severely subnormal who live in their own homes; the best methods of care for those who cannot be looked after at home; and methods of training imbeciles for sheltered employment.

1. The Severely Subnormal at Home

Saenger, in an important monograph (1957), reports on the investigation of a large sample of medium to high-grade imbeciles aged between 17 and 40, resident in New York. Slightly more than half of the very large sample appeared to be outspokenly happy and contented; neurotic trends were suspected in 20 per cent and psychotic symptoms in 6 per cent. Three-quarters showed secondary physical handicaps, 20 per cent disturbances of motor co-ordination and 40 per cent suffered from speech defects. Only one out of every 9 managed to read even a simple passage. They thus presented a picture of considerable impairment.

Saenger writes that the majority had made a good adjustment to their homes. One in 4 participated in everyday family life: they took an active interest in the affairs of other family members and tried to help within their limitations. Roughly half could communicate in a limited way with other members of their family without developing a genuine give-and-take relationship. An almost vegetative kind of existence, with mechanical obedience to commands, characterised the remaining cases.

Three-quarters of the parents felt that their child was easy to get on with and presented no major difficulties. Problems

which did exist were those of the normal 5-6-year-old child: restlessness, temper tantrums and obstinacy were the commonest. Only 5 per cent of parents felt that their child was difficult to handle. As with normal children, the main feature of families where imbeciles showed major behaviour problems was family tension and rejection of the child. In general, the principles of mental hygiene known to operate in the normal population applied to this group.

Adjustment to the community obviously presents greater difficulties than adjustment to the home. Most of these persons spent the greater part of the day at home, but nearly 80 per cent went out alone, at least occasionally. Only one-third, however, were able to leave the immediate neighbourhood and travel unaccompanied by bus or tube train. Community adjustment would clearly be easier in a rural area. On the whole, these defectives were relatively solitary and friendless and the majority showed no interest in members of the opposite sex. Very few—only 7 per cent—had got into trouble with the authorities and the offences were of a minor order.

Twenty-seven per cent of the sample were working for pay, and an additional 9 per cent had had previous employment but at the time of the study were out of a job. The work involved simple domestic tasks outside the home—sweeping, washing up, or unskilled labour in shops or factories.

The picture presented by this transatlantic study of social factors in low-grade deficiency accords well with the point of view developing from English research. It is interesting that probably no vocational training was given to these American imbeciles; if it had been, it seems exceedingly likely that the position might have been even better.

In this country Tizard and Grad (1961) studied the families of 250 idiots and

imbeciles living in the County of London; some of these children were at home and some were in institutions. The latter tended to be those with severe problems but by no means exclusively so. It was found that the families with a child at home were worst off in terms of housing, over-crowding and finance, and that those whose children were young or most severely handicapped suffered the greatest hardships. These authors summarise the findings of this survey, and discuss the implications.

2. Imbeciles in Institutions

There are 8,500 children in mental deficiency institutions in England and Wales, mostly of imbecile or idiot grade, and something like 25,000 low-grade adults. Since it seems likely that this is a growing problem, the effect of housing youngsters in large colonies has been the subject of researches by Lyle (1959) and Tizard (1960). A controlled comparison of imbecile children housed in a small, specially staffed unit, with a group in the parent hospital, suggested that long residence in an institution retards verbal intelligence, a point which in this specific field is fully consistent with findings at a higher intellectual level. Furthermore, the most striking effect of the ordered but free regimen provided in the small unit was the desirable effect on the children's social and emotional behaviour. Starting as aggressive, destructive and intolerant of frustration, they appeared after some months to have become more tractable, co-operative and generally adjusted. Tizard believes that this is due to the greater individual attention available in his unit, and to the special educational programme which differed from that generally adopted in occupation centres for mentally handicapped children. However, we must note in passing that some institutions have solved many of these problems simply by

increasing their staffing, and allotting staff permanently to individual children in the ratio of 1:3 (Klackenberg 1956).

3. Education and Training of Imbeciles

There are two main sources of evidence: on the one hand, the experience of occupation centres and those E.S.N. schools who cater for classes of the severely subnormal; on the other hand, experimental studies conducted in workshops and laboratories, which aim to examine in rigorously controlled conditions factors underlying imbecile learning and trainability. Regrettably, the wisdom and knowledge gained in the former circumstances are less frequently recorded than in the latter, and, furthermore, provision for daily occupation in the community is a relatively new concept, which has not yet come to full fruition. There is, however, increasing acceptance of the idea that imbeciles, like normal young people, need to 'go to school' or 'out to work', to be part of a community adapted to their needs and to be subjected to a type of discipline which can only be provided in a social setting. It has also been suggested that weekly residential centres might meet the need of families whose imbecile children are exceptionally disturbed, or where financial circumstances are such that the mother should work.

Another problem is the great dearth of properly trained staff to cope with the expansion of occupation services for the severely subnormal and recently the Ministry of Health has set up a Committee to inquire into this matter.

So far as experimental studies of imbeciles are concerned, fewer investigations have been carried out on children than on adults. Examples of recent work in each sphere will be presented.

(a) Children

A series of experiments on the formation

of verbal concepts in imbeciles have been conducted by Hermelin and O'Connor, and some of these have been described in an article in this Bulletin (1959). In one study (1958) they observed a group of children aged between 10 and 16. Using discrimination tasks, some of which could only be learned by rote while others allowed the possibility of solution by application of a generalising principle, these authors found that the imbeciles made effective use of the simple classifying concepts which they had spontaneously developed.

More recent unpublished work by these authors concerns the relative effectiveness of cross modality versus like modality learning in imbecile children. The identification of pictures from among a larger group of similar pictorial objects or the recognition of spoken nouns from among similar nouns formed the basis of experiments in like modalities. Cross modality recognition involved either the learning of auditory signals and the identification of their visual equivalents or vice versa. This latter was found to be the more effective method of learning. Older children appeared to have better powers of recognition or alternatively the younger ones learned more slowly. A further finding was that whereas the visual discrimination of imbecile adults was inferior to that of normal controls of the same mental ages, and significantly inferior to that of normals of equal chronological age, their stereognostic ability was equal to that of controls of similar chronological age.

The effects of pre-school stimulation of mentally retarded children have been investigated by Kirk (1958) who included a small percentage of imbeciles in his group. He found that in the main such children increased their social and intellectual growth rates compared with controls who had not enjoyed special pre-school facilities. But, although this increased rate

was maintained during the first period of school life, others caught up after a few years of schooling. This suggests that limits had not been fundamentally altered but that the children had merely reached them earlier.

Clarke and Blakemore (1961) suggested that the younger the imbecile the more likely he was to be able to generalise his training experience, that is, as psychologists put it, to show 'transfer of training'. Using four relatively difficult perceptual-motor tasks, each of which was composed of two forms, their hypothesis was strongly confirmed, for imbecile children averaging nine years of age showed far greater transfer than seventeen- and twenty-three-year olds, whose learning curves were very similar. Moreover, the authors found that with practice the children successively approximated to the performances of adolescents and adults, so that after 20 trials on each type of task they had nearly achieved the end-levels of the older subjects. In other words, nine-year-old imbeciles functioned nearly as well as the adults. The authors suggested that two things were transferred: first, the mental 'set' or preparedness to do the particular type of task, and second, a sharpened perceptual and conceptual discrimination.

Subsequent experiments, as yet unpublished, indicate retention of learning over long periods of non-practice, and the facilitatory effect of early learning upon the subsequent acquisition of perceptual-motor skills. It seems clear that the usual methods of training imbecile children do little to alter slowness in learning and perceptual handicaps. Yet it is equally clear that in both spheres these deficits can be partly overcome by relatively simple methods founded on the ideas which Itard originally propounded 150 years ago. Moreover, it is clear that in the training of persons of this nature there has been too great a readiness, no doubt for entirely

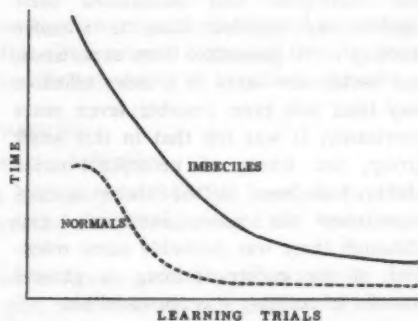
humane reasons, to accept the imbecile's handicap rather than to help him overcome it. The result has often been reinforcement of inherent weaknesses, particularly with regard to lack of speed in various spheres of activity.

All these findings appear to add further justification to the statement of Clarke and Hermelin (1955) that 'qualities such as manual dexterity and motor co-ordination are not static but capable of improvement within limits which are often ill understood and ill defined'. Once again, however, we do not know whether approaches such as this alter fundamentally the upper limits of development in imbeciles, or whether such limits are merely achieved earlier. Even, however, if it is only the latter it is of great theoretical and practical significance, and suggests that the earlier the training the more likely it will be that the handicap may be minimised.

(b) *Adolescents and Adults*

Traditionally, clinical opinion of the imbecile's trainability has been exceedingly gloomy—a general principle which seems to recur in the history of almost any type of handicapped persons. Yet in other spheres, some modification of the earlier pessimism has been justified. And so, too, with imbeciles. Two articles in this Bulletin (Clarke 1959 and O'Connor 1959) have reviewed some of the studies of recent years, and have shown that clinical opinion, emphasising what the imbecile *cannot* do, has in general been correct only in so far as short-term observation or training is concerned. Limitations on learning, though considerable, are however by no means so profound as had earlier been thought. The main feature is learning slowness and a wide variety of low levels at the start of any learning task. In general, however, individual differences between imbeciles get smaller and smaller as learning proceeds and all tend to end at

very similar points, often not grossly inferior to normal persons. The curves shown in the figure illustrate the latter principle.



Typical learning curves for imbeciles and normals on simple tasks.

A series of laboratory and workshop studies has indicated that most able-bodied imbeciles are capable of acquiring a variety of simple industrial skills, working in sheltered conditions, provided they are well motivated and taught according to certain specific principles. Operations involving dexterous manual movements, such as folding and glueing card-board boxes, packing objects in cartons, simple assembly and soldering have been successfully acquired. In addition, a variety of sorting tasks involving visual discrimination have been found appropriate, and imbeciles can be gainfully employed at standard piece-rates doing a number of jobs essential to light industry. Furthermore, there is evidence to indicate that such training is not only specific but generalises (or transfers) to other tasks. Thus, recently, Clarke, Cookson and Clarke (to be published) investigated the effects of seven years' special stimulation on a group of adult medium to low-grade imbeciles in a sheltered workshop. Re-testing in 1959 on a spatial relations test, used by Tizard in 1952 as a learning task, they found a greatly enhanced performance.

There were two possible explanations: first, that the imbeciles had remembered the task, and second, that the variety of industrial experience which the imbeciles had undergone had diminished their rigidity and enabled them to transfer training or to generalise from experience, and tackle new tasks in a more effective way than had been possible seven years previously. It was felt that in this adult group, the limits of perceptual-motor ability had been shifted above normal expectancy for adult imbeciles, and although there was probably some retention of the earlier training, a general transfer of training was also possible.

Conclusions

The severely subnormal were the last major section of the gravely handicapped

to attract occupational and psychological research, and it is only during the last decade that such investigations have been undertaken in any quantity. Against the background of a considerable increase in their numbers, it has already been found that something can be done to minimise some aspects of their handicap. As in other fields, there has been a fruitful interaction between theory and practice, with each modifying and challenging the other. Above all it is clear that in the task of finding more about the behaviour and more about ways of helping imbeciles to a fuller life nothing can replace careful, controlled experimentation in which the pedestrian, but rewarding, methods of science are used. We have learned a little about this field—there is a great deal more useful knowledge to be discovered.

SUMMARY

This article attempts to give a brief global picture of current psychological research in the investigation and modification of imbecile behaviour.

Starting with the assumption that an increasing number of congenitally severely handicapped individuals are surviving, attention is focused on three main areas of research into their social adjustment:

- (1) the severely subnormal in their own homes;
- (2) the problem of the best methods of caring for those who must be removed to institutions;
- (3) the education and training of imbeciles, viewed from the point of view (a) of occupation and training centres and (b) of experimental investigations in more controlled conditions.

It is concluded that this is a rewarding field of investigation in which only a minimal amount of established fact is available. Already, however, it appears that considerably more can be done for the severely subnormal than formerly thought.

RÉSUMÉ

Etude du comportement de l'imbécile

On trouvera dans cet article un tableau bref et global tendant à présenter les découvertes psychologiques actuelles relatives à l'étude et aux modifications du comportement de l'imbécile.

Tenant compte du fait qu'il y a de plus en plus de survivants parmi les êtres atteints d'infirmités congénitales graves, l'auteur concentre son attention sur trois principaux secteurs de recherches touchant à leur adaptation sociale:

- (1) les grands déficients mentaux dans leur famille;

- (2) le problème des meilleurs méthodes de soins à appliquer à ceux qui doivent être placés dans des institutions;
- (3) l'éducation et la formation des imbéciles considérées du double point de vue (a) des centres d'occupation et de formation; (b) des recherches expérimentales dans des conditions plus rigoureuses.

L'auteur conclut qu'il s'agit d'un terrain d'investigation 'payant' pour lequel cependant on ne dispose que de très peu de données. Toutefois, dès à présent, il apparaît que l'on peut faire bien davantage pour les enfants gravement déficients qu'on ne le pensait naguère.

ZUSAMMENFASSUNG

Studium des Verhaltens des Imbezillen

In diesem Artikel wird versucht, ein kurzes und globales Bild der heutigen psychologischen Forschungen über die Untersuchungen und die Veränderungen des Verhaltens des Imbezillen zu geben.

Der Autor geht von der Voraussetzung ab, dass es immer mehr Überlebende unter den Kranken mit angeborenen schweren Defekten gibt, und lenkt seine Aufmerksamkeit auf drei hauptsächlichste Forschungsgebiete, betreffs ihrer sozialen Anpassung:

- (1) Die Kranken mit schweren Schwachsinnformen in ihrer Familie.
- (2) Das Problem der besten Pflegemethoden für diejenigen, die in Anstalten untergebracht werden müssen.
- (3) Die Erziehung und Ausbildung der Imbezille vom Gesichtspunkt (a) der Beschäftigungs- und Ausbildungszentren, (b) der experimentellen Untersuchungen in den genauesten Bedingungen.

Der Autor schliesst, dass hier ein lohnendes Forschungsgebiet ist, in dem man aber nur über sehr wenige feste Tatsachen verfügt. Jedoch, schon jetzt erscheint es, dass man viel mehr für die schwer abnormen Kinder tun kann, als man früher glaubte.

REFERENCES

- Benda, C. E. (1952) *Developmental Disorders of Mentation and Cerebral Palsies*. New York: Grune & Stratton.
- Carter, C. O. (1958) 'A life-table for mongols with the causes of death.' *J. ment. Defic. Res.*, 2, 64.
- Clarke, A. D. B., Blakemore, C. (1961) 'Age and perceptual-motor transfer in imbeciles.' *Brit. J. Psychol.* 52, 125-131.
- Hermelin, B. (1955) 'Adult imbeciles; their abilities and trainability.' *Lancet*, ii, 337-9.
- Clarke, A. M. (1959) 'Teaching imbeciles industrial skills.' *Cer. Palsy Bull.*, 1, No. 6, 14-18.
- Hermelin, B., O'Connor, N. (1958) 'The rote and concept learning of imbeciles.' *J. ment. Defic. Res.*, 2, 21-7.
- Kirk, S. A. (1958) *Early Education of the Mentally Retarded*. Urbana: Univ. Illinois Press.
- Klackenberg, G. (1956) 'Studies in maternal deprivation in infants' homes.' *Acta Paediatrica*, 45, 1-12.
- Lyle, J. G. (1959) 'The effect of an institution environment upon the verbal development of imbecile children.' *J. ment. Defic. Res.*, 3, 122-8.
- O'Connor, N. (1959) 'Problem solving and mental defect.' *Cer. Palsy Bull.*, 1, No. 6, 9-13.
- Saenger, G. (1957) 'The adjustment of severely retarded adults in the community.' Report to the New York Interdepartmental Health Resources Board.
- Tizard, J. (1960) 'Residential care of mentally handicapped children.' *Brit. med. J.*, i, 1041-6.
- Grad, J. C. (1961). *The Mentally Handicapped and their Families*. London: Maudsley Monographs, Oxford University Press.

Use and Abuse of the Film in Recording the Behaviour and Reactions of the Newborn Infant*

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CASE histories and the verbal description of a patient's condition occupy a time-honoured place in the practice of medicine. However, the writing of such notes can be time consuming, especially if it is planned to study large numbers of children over a protracted period of time. A more serious handicap is perhaps the chance of misinterpretation. Case-notes represent the observations and views of one observer, and his interpretation of the facts is all that is normally available for further study and comparison. Thus inaccuracies and misinterpretations may well arise due to differences in language, terminology and medical background, to mention only a few. Furthermore, it is not an easy exercise to draw conclusions from the written notes of one child at different ages or from the records of different children at the same age.

A pictorial record, however, if well designed and executed with due regard to standardisation, is a permanent document of facts which can be evaluated as primary evidence without the handicap of language and other barriers. Equally, pictures can store the available information in a greatly reduced space and readily lend themselves to comparison with other, related pictures.

* Read at a Study Group on the Neurological Examination of the Newborn, Groningen, 1960.

Motion Picture Film

A single photographic print, unless it contains a complex of images, can only depict one single static image for the study of voluntary and involuntary movements. The motion picture film can provide a continuous record of movement by dissecting it into separate phases which, on projection, are linked again by the persistence of visual images on the retina.

Before discussing the possible application and techniques of the film, it is desirable to establish the limitations. The film can record movement and it can recreate events or phenomena of the past, so that it can act as an aide mémoire or as a visual memory of behaviour and reaction. Here also lies its limitation: while it may be less distracting to observe movement on the projection screen in a darkened room and, as it were, through a window, the mind is still incapable of rational interpretation before one image has given way to the next and one phase of movement has been replaced by another. The inevitable conclusion is that the film can at best yield only a subjective impression. Even so, the film may come to be regarded as the best means at our disposal of comparing the development of one child at different ages or the behaviour

and reaction of different children at the same age (Fig. 1).

Standardisation

This horizontal and vertical comparison will gain in validity in direct relation to the degree of standardisation which can be brought to bear on the preparation of the film records.

Insufficient attention has been paid to standardisation in scientific recording; this is particularly true of medical and scientific photography which have otherwise reached a high level of perfection and usefulness. If one man is working for his own end it is relatively easy for him to develop a discipline which will result in uniformity of results whatever form these may take. However, when more than one recorder is involved, or where there is a need to compare results of one observation with another, or of one hospital or centre with another, difficulties immediately arise.

Standardisation in motion picture recording offers special problems and,

by the very nature of the medium, the film does not lend itself to such hard and fast rules as still photography. The problem as a whole is of such fundamental importance in medical photography that it may be discussed at practically any level of complexity. It will be considered here extremely broadly as to how best to harness cinematography to recording the newborn infant.

Apparatus

Briefly speaking, standardised recording hinges on area (or scale), viewpoint, lighting, tone and contrast being held constant from one record to another in a series. In cinematography we have to add to this the rate of recording—and subsequent projection. Strict attention must be paid to all these points at the time of actually making the film record, particularly since there is little or no opportunity to make minor adjustments to the finished film. This is in complete contrast to the conditions which obtain in still photography.

CHILD	1 day	1 month	6 months	1 year	2 years	5 years
1						
2						
3						
4						
5						

Fig. 1. Standardised film records may be used for *horizontal* comparison of one child at different ages, and for *vertical* comparison of different children at the same age.

It is simple to standardise camera equipment, once the decision to use 8 or 16 mm. has been made. Lenses are supplied in standard focal lengths, and suitable choice will be discussed later. The choice of the film type and lighting must also be considered. Variable rate of recording requires a camera with speed control. This is also important in relation to the addition of sound, which will be dealt with separately. The proper choice of background is a subject in itself; here it is probably sufficient to say that they should be devoid of distracting features and should be of an even neutral tone, black or white probably being preferable to grey, possibly some form of measuring grid should be included, though these can be misleading. An alternative is to fix absolutely the camera subject distance and to fit some form of graticule in the actual camera gate.

Methods

The first cardinal rule in standardised recording is that the position of the camera must be fixed in relation to the subject, not only in terms of distance, but also angle or viewpoint to maintain a comparable perspective or viewing angle. Changes of scale should be achieved by varying the focal length of the lens rather than by moving the camera. The optimum position of the baby in relation to the camera is more difficult to determine, but a three-quarter lateral look-down position would seem reasonable (Fig. 2). Routine records might start with a few feet showing the behaviour of the baby without external interference. Thereafter a sequence of handling or examination must be evolved and adhered to on each occasion.

Naturally it may be necessary to establish more than one viewpoint but this merely adds to the complexity of the procedure. However, it is not implied that the camera need be fixed to a tripod,



Fig. 2. Suggested standard 45° look-down position for filming examination. Note grid markings on table.

providing there is some method of accurately establishing camera to subject distances which is all important.

The length of each shot has a particular significance not only from the point of view of subsequent viewing, but, apart from electrically driven cameras, there is a limit to the length of time which the motor will run. It is probably better to have a number of brief shots each long enough to communicate what is going on, which can be repeated once if necessary. It is well to remember that duplicate prints can always be obtained subsequently and strung together for more prolonged viewing or analysis; the film loop for continuous showing may also have an application here.

Finally, an identity number should be included in the picture area. This is too obvious to warrant elaboration, but, clearly, newborn babies are very much alike, and this simple measure saves an enormous amount of later sorting out and is important in general documentation. Cloakroom tickets are admirable for this purpose. It is essential that the operator takes complete notes about each record, making dimensioned sketches of the set-up where this is unusual. He must at least

make a note of the camera subject distance, the focal length of the lens, the lighting and the rate of recording.

Recording

A practical method of obtaining clinical motion picture records with very simple equipment may well be the approach needed for the problem of recording the behaviour and reactions of the newborn infant.

Equipment

Full technical details of a system involving a magazine camera which is hand-held and carries its own lighting have already been published (Hansell, P. 1957).

Nowadays the zoom or variable focus lens has come into its own and it offers enormous advantages. These lenses are still very expensive but they do enable one to eliminate a range of individual lenses which might otherwise be required for comprehensive work. In this context the cheapest form of camera body is treated merely as a film magazine to attach to a zoom lens with the additional facility of through-the-lens viewing and focusing.

Permanent Installation

The concept of a photographic observation ward was first formulated by R. C. Mac Keith a number of years ago. Automatic or semi-automatic filming may be deemed desirable to achieve standardisation and thus comparability, and also when it is necessary to record without the knowledge of the subject to exclude variable factors in behaviour.

A permanent arrangement can consist of a hidden camera which is either remote controlled at will or which can be activated by a timing device so that a given length of film is exposed at regular intervals. Correct exposure can be ensured either by excluding all daylight and using a constant

quantity of artificial illumination, or by fitting an automatic iris control, which will adjust the lens diaphragm according to the amount of light falling on a photo-electric cell. Unless the camera can be reached from another room, the changing of lens and focusing present a problem. Such an arrangement would normally be restricted to one view point, one angle of view and a fixed distance between subject and camera.

It may well be deemed an advantage to examine and record the newborn in a room which is specially equipped for this purpose. As the filming may have to be done at any time of the day or night, it would be impracticable to make one person responsible for the camera, so that the equipment ought to be as automatic as possible to achieve the desired degree of standardisation.

Suggested Scheme

A possible solution consists of a camera fixed on an arc, so that it can travel from a position vertically above the infant to a point where it is parallel to and just above the level of the examination table. The table is pivoted, so that the baby can be rotated in front of the camera for lateral

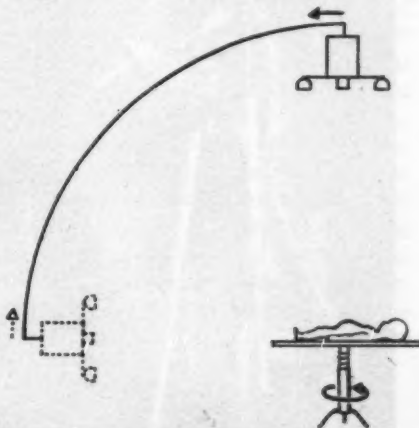


Fig. 3. Standardised filming with the aid of a television camera working at predetermined angles and distances.

views. Constant illumination is achieved by fitting a lamp on either side of the camera. A zoom lens is used to obtain any desired degree of image reduction. (Fig 3)

If the conventional film camera is replaced with a vidicon television camera, focusing and adjustment of the zoom lens can be carried out by remote control, until the image on a television receiver is sharp and shows the scale and view point laid down for standard recording. A film camera can then be activated to record from a second television receiver. One physician can thus examine and record a newborn baby under standard conditions single-handed. (Fig. 4)

A normal film camera can, with little adjustment, be used in conjunction with a synchronising unit for television recording. However, only every second sweep or scan will be recorded, so that the vertical definition of the image is bound to suffer. Cameras with fast pull-down mechanism

are now available which can record every scan and thereby double vertical definition, although capital expenditure may rise by approximately £500. To avoid the expense of television recording, a film camera may be linked in all respects in parallel with the television camera.

This scheme relies on almost complete automation, and it is essential to record a notice board which carries the essential data about the infant, before the baby itself is placed on the examination table. This will avoid possible confusion after a film has been processed and is separated for storage as individual records.

Simultaneous Recording of Other Evidence

The results of electro-cardiography, -myography or -encephalography may also be recorded on film. It may be important to see the state of the patient and at the same time appreciate the normality and abnormality of electrical impulse patterns.



Fig. 4. Experimental arrangement showing baby, T.V. camera, monitor for the physician and second monitor with film camera attached.

Essentially there are two methods of incorporating other visual evidence of this type in a motion picture film. One is the split frame technique whereby only half the width of the film is exposed on the first subject; the film is then wound back and re-exposed on the other material so that two different images appear on the screen side by side. This technique may have its application in certain instances, but as only one camera is used the two phenomena cannot be recorded at one and the same time. When this is important, there is no alternative to using two or more cameras running at the same time but recording different phenomena. The essential factor is that the records must be synchronised by some visible signal which will act as an index mark on each individual record when it has been processed; it then remains a laboratory procedure to have, for instance, two strips of film optically reduced and printed side by side in synchronism on the film which is to be examined. Without going into this rather specialised technique very deeply, a short length of film can combine three distinct types of evidence in one record. In visual form the posture of the patient and the appropriate electro-myographic traces on the cathode-ray oscilloscope are shown, and the audible signals on a synchronised sound track are added.

Sound

A sound track may be added for different reasons, either as purely informative or narrative after the film has been put together, or to communicate additional evidence in the form of naturally occurring or artificially produced sounds. In this instance it is recorded on disc or tape in synchronism with the picture material and later transferred to the actual film itself.

Both these types of track are applicable to the problem of recording the newborn. In the first example a commentator's voice

recording wild (not synchronised with the images appearing on the screen) could be of value in a master film to indicate the particular sequence of examination to be adopted at one or more centres. This would be termed instructional and would be aimed at standardisation of method. A use for the second type of track might be found in the addition of heart sounds or the child's cry to be recorded in synchronism with the picture.

It is therefore necessary to decide first of all whether sound is necessary for one or other of these reasons and then to investigate the means.

The various methods are:

1. Synchronised track recording on location.
2. Simultaneous recording in the camera.
3. Tape recorder—interlocked or free.
4. Play back as film and tape separately.
5. Play back as stripe (magnetic) on film (4 types).
6. Play back as film with optical track.

A fairly compact and reasonably inexpensive installation, which is used for making sound tracks of various types as well as for normal projection in a large auditorium of films which carry every known type of sound track, may be cited to illustrate the application of these methods (Fig. 5). The complex machinery of the sound recording side should not concern us very greatly, but it is well to remember that nothing is lost by having a quite ordinary tape recorder handy as part of the general recording apparatus. It may or may not be found necessary to damp the noise of the camera or to use a highly directional microphone which does not pick up background noises.

Film Records

No scientific film should ever be projected in its original form. It has recorded



Fig. 5. Typical domestic installation for recording and playback of all types of magnetic sound track (Siemens). The commentator, in a sound-treated booth, views the small picture in front of him. Film is run on the right-hand side and master tape on the left.

an event which cannot be repeated, so that the master must be protected from possible damage. All records should therefore be copied as a matter of course.

Each copy should then be filed in a separate box, labelled with the code number of the baby and a letter of the alphabet to denote whether it was the first, second or third film of this infant (e.g. 126A, 126B, etc.). This will make it possible to compare any one record with any other of the same child or with that of another child.

The same code system should also be used on the envelope which contains any written or magnetic sound comments on the individual film records. The outside of the envelope should show the code number and the letters with related date and age at the time of each recording. Films and envelopes can then be filed in numerical order, so that the punch card system used for storing other information can be used to locate any one of the films and its associated data.

Projection

If standard image reduction and standard

speeds have been used during recording, it is naturally important to project the resultant films at a standard magnification and at a standard speed. Unless a grid on a transparent base was used in the gate of the camera, or a gridded background under and behind the baby, it may help to project on to a gridded screen.

Although two films can be compared by projection in sequence, it may be thought worthwhile to link two projectors in parallel so that two films can be shown side by side simultaneously. As each movement may require repeated study, endless loops may be more convenient. While most projectors can be adapted for loop films, the Lytax projector has been designed to accept loop film magazines which can be clipped on or removed while the apparatus is running. The Institute for the Scientific Film in Göttingen stimulated this development with their design of a projector which makes it possible to switch from one loop film magazine to another in rapid succession.

To reduce the speed with which images pass across the screen it is possible to use special projectors for single frame, or two

frames per second, projection. Yet other projectors can be controlled from a distance for screening at variable speeds both forward and in reverse.

Detailed study of individual frames, for the tracing of specific movements from picture to picture, can be carried out with special equipment. A foot switch controls the advance from one picture to the next. A static image is projected on to paper so that a complete tracing or graph can be constructed. For approximately £8,000 an American machine can be purchased where a pointer aimed at the part of the image to be analysed will direct the production of a punched tape which is then fed into an automatic analyser.

As children reach an age when they can learn to perform simple tasks, for instance to reach for a toy, it may be possible to reduce the recorded movement to a more or less simple graph which is readily compared with other, similar graphs. Still cameras can be employed for this purpose,

using a larger image format but a reduced rate of recording with a proportional saving of film. While the film is advanced by a pre-set timing mechanism, any desired shutter speed can be applied, so that each picture is perfectly sharp. This method lends itself to tracing particular features on to a single sheet of translucent graph paper. As successive pictures are projected on to the back of the graph paper, a static reference line, common to all images, is brought into register with the tracing of this line from the first picture. One or more points on a moving limb are then traced through. The resulting graph will be an interpretation of movement in two dimensions measured against time, as the interval between images is a known quantity. While full three-dimensional evaluation would require two synchronised cameras at right angles to each other, this and other limitations may yet be outweighed by the ease with which these graphs can be compared and stored.

SUMMARY

The apparatus, methods and evaluation for film recording are outlined, with particular emphasis on standardisation for comparability. Questions of finance and administration must be decided in relation to other requirements. A comprehensive programme of standardised visual recording promises to contribute to the establishment of what constitutes a normal newborn. From this it may be possible to diagnose abnormalities at a very much earlier age than at present, and to assess the effectiveness of treatment and education with enhanced accuracy.

RÉSUMÉ

Usage et abus de l'enregistrement filmé du comportement et des réactions du nouveau-né

Appareils, méthodes et évaluations sont décrits avec référence particulière à la normalisation dans un but de comparaison. Il s'agit de trancher de questions financières et administratives par rapport à d'autres besoins dans le cadre général de la conférence. Cependant un programme rationnel d'enregistrement visuel permet de contribuer à l'établissement de ce qui constitue un nouveau-né normal. Cette méthode permettra de diagnostiquer les anomalies à un âge beaucoup plus précoce qu'aujourd'hui et d'apprécier l'efficacité du traitement et de l'éducation avec une précision accrue.

ZUSAMMENFASSUNG

Gebrauch und Missbrauch des Films für die Wiedergabe des Benehmens und der Reaktionen des Neugeborenen

Apparate, Methoden und Bestimmungen werden beschrieben, mit besonderer Rücksicht auf Vergleiche gestattende Normierung. Finanz- und Verwaltungsfragen müssen in

Verbindung mit anderen Forderungen im allgemeinen Rahmen der Konferenz gelöst werden. Ein umfassendes Programm für normierte Wiedergabe erlaubt es jedoch, zur Festsetzung der Züge eines normalen Neugeborenen beizutragen. Es wird dadurch möglich werden, Abnormitäten in einem viel früheren Alter als gegenwärtig zu erkennen und die Wirksamkeit der Behandlung und Erziehung mit erhöhter Genauigkeit abzuschätzen.

REFERENCE

- Hansell, P. (1957). 'Simplified cinematography., *Britt. J. Photogr.* 104, 208.

Risk Register

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There Methods of Detecting Defects in Children

IT is common knowledge to all who have contact with Cerebral Palsy, Audiology, Ophthalmic, and other allied Paediatric Units, that far too many children with diagnosable defects are still finding their way to these Units for expert evaluation only after irremediable damage has occurred, and at far too late a stage for effective rather than palliative treatment to be carried out. This is partly due to the fact that detection has always been, and still is, mainly based on the appearance of symptoms and signs obvious enough for parents to notice. This point of time in itself is usually too late for optimum treatment and even so is then followed by an interval during which the parents make up their minds whether or not to seek advice from their doctor, and often yet another interval before the child is seen by an expert team.

Two other methods of detection stand out as ways of mending this often tragic situation: First, the country-wide screening of the total population of infants, without exception, area by area, by panels of experts, several times in the first year of life, or until the panels were satisfied that all was well. Secondly, the screening of just those infants who have undergone some pre-natal, natal or post-natal insult or risk known to predispose to the development of the defects in question; this technique in relation to the detection of hearing defects in a selected group was described by Fisch (1955), and will be elaborated later.

Of the three methods mentioned—the symptomatic, the total population screening, and the screening of the ‘at risk’ group—the second appears to be the ideal one, but for many reasons is, in present conditions, unattainable. Also, the frequent detailed examination and re-examination of healthy babies without definite indication may be of questionable wisdom from both the child’s and the parents’ point of view, and possibly also from that of the examiner’s continued keenness and enthusiasm. The factors of sheer numbers, of time loss and inconvenience to parents, of time consumed and availability of expert medical personnel, and the prohibitive financial implications of such a scheme must be balanced against the return in the earlier detection of handicaps in a remediable stage. To cite one of the several surveys on this issue, Howorth (1958), investigating the subject of early detection of deafness, attempted, with the aid of selected trained health visitors, to test all preschool children in an area and found 2 cases with significant hearing loss in 3,065 examinations, which is the equivalent of 0.65 per 1,000; whereas when 662 cases of ‘at risk’ children were tested, 6 cases were found, which is the equivalent of 9.06 per 1,000.

It seems clear from this and similar investigations that the method of concentrating on the considerably smaller group of ‘at risk’ children (who, on most existing registers, amount to around 20 per cent of the total child population) is a more practical approach, and certainly an advance, with notable exceptions, on

the present generally haphazard way that defects are eventually detected.

When there are enough otologists experienced and expert in screening infants, and sufficiently equipped premises for them to work in, the figure of 9.06 per 1,000 quoted above will probably be significantly exceeded in cases of hearing loss detected.

Most cases of deafness arise later in life, but, from the point of view of speech and communication, it is the early diagnosis of the congenital and infantile deafness that is important. In this context Harrison (1960) and Bordley (1960) said they had been unable to detect the cause of deafness in 30 per cent and 22 per cent of their cases respectively. In the case of congenital deafness, however, the aetiology is unknown in some 40 per cent of cases (L. Fisch, personal communication).

In the case of cerebral palsy, Professor Illingworth (1960) said that 'early recognition of cases of cerebral palsy depends partly on following up infants who have been exposed to special risk, but also on a very close knowledge of development'. Mitchell (1959) states that, whereas prematurity, abnormal birth, or postnatal disease provide a possible explanation for all but 14 per cent of cases of cerebral diplegia, the factors were absent in 38.2 per cent of cases of hemiplegia. In a British survey of 385 cases of cerebral palsy, including all its subdivisions, Woods (1958) found that 35 per cent were premature, 21 per cent followed an abnormal birth, and a further 28 per cent had severe neonatal symptoms following a normal birth; thus 84 per cent showed a clear 'risk' history.

In the case of eye defects the approximate percentage figures are not available, but Drillien (1958) found that 65 per cent of children with a birth-weight of under 4 lb. had a visual defect by school age, the commonest single lesion being myopia. Gardiner (1960) showed that 75 per cent

of children born myopic came into the world after a pregnancy disturbed by illness, a rate about four times as high as a control group.

In view of the above findings (Gardiner, personal communication and 1961) recommends refracting all children at the age of 2 years who were born under 4 lb. in weight, whether or not they have visual symptoms.

Broadly speaking, therefore, roughly 70 per cent of defects can be traced at a treatable stage if all resources are concentrated on 20 per cent of the infant population, so that this approach promises to be well worth while. As for the remaining 30 per cent, so long as their cause remains obscure and specialist evaluation of the total infant population is unattainable, they must continue to be detected, probably rather later, by the existing 'symptomatic' method—e.g., by the recognition of delay in arrival at milestones and appearance of signs and symptoms, etc.

The symptomatic method has great disadvantages to parents as well as to the affected children. Parents will often say they had suspected for some time that something was amiss, but had not been sure enough to seek advice. This uncertainty can, and does, cause an anxiety bordering on panic, and a distress which is almost unendurable. This mental suffering dulls the parents' thought processes and must further affect their relationship with the child; it can be avoided or shortened, or even shown to be unwarranted, by early and, most important, expert advice. Furthermore, if they are given the necessary knowledge regarding the challenge they have to face, most parents will unflinchingly accept and deal efficiently and devotedly with the most difficult problems.

Disorders Detectable in Infancy

In present circumstances the following

conditions lend themselves most readily to earlier recognition by means of a 'Risk Register'. A few reasons for their inclusion are briefly given:

A. Cerebral Palsy: (1) Full early medical assessment, including the evaluation of motor capacity, hearing, vision, sensation, capacity for speech, intelligence and personality.

(2) Psychological support for the parents, giving them the vital feeling that they are doing something and that everything necessary is being done for the child. Promotion of good relationships between the parents, siblings and the handicapped child from an early date, and social training in a nursery if necessary.

(3) The management of the infant's movements from an early and plastic stage by the physiotherapist, and by the parents under the physiotherapist's guidance.

B. Hearing Defects: Early diagnosis and treatment, preferably by six months of age is now recognised as of vital importance for the subsequent development of language and speech. Treatment consisting of the fitting of hearing aids as necessary, guidance of parents in appropriate management and training of the child, and auditory training by an expert team.

C. Visual Defects: Early and rapid diagnosis of defects and expert treatment of conditions such as latent and manifest squints in order to promote binocular vision and avoid the onset of suppression amblyopia.

Wellesley Cole (1959) stated that 'out of every 100 children, between 5 and 6 are destined to lose the use of one eye' and goes on to say, 'in a country with the most comprehensive Health Service in the world it is paradoxical that its young children should be allowed to run the risk of suffering this grave disability through neglect'.

D. Other Conditions which can be included on a 'Risk Register' are children

born into families with histories of familial, hereditary and other congenital disorders such as muscular dystrophies, amyotonia congenita, haemophilia, Friedreich's ataxia, and phenylketonuria. Early recognition may enable treatment of the child and support for the family as a whole. Referral to a Genetic Unit may prevent further tragedies or remove unnecessary anxiety in the parents. More research is needed into conditions that could be detected or prevented in this way.

Aetiological Categories for a Register

There is no space here for an exhaustive survey of categories that could be included on a Risk Register, and the following skeleton classification can be considerably added to or re-arranged.

A. RISK CATEGORIES

1. Hereditary:

Those who have a family history of the conditions to be detected or if possible prevented, such as cerebral palsy or other CNS defect, epilepsy, deafness, squints, translocation mongolism, mental deficiency, etc. Also the children of unmarried mothers, for they have a higher incidence of defects.

2. Prenatal:

Rubella or other severe virus disease in first 16 weeks of pregnancy, to exclude cerebral palsy, deafness, and eye or cardiac defects.

Threatened abortion or any antepartum haemorrhage.

Severe illness, accident or operation in first 16 weeks of pregnancy, especially if anaesthesia was necessary.

Hyperemesis.

Toxaemia.

Diabetes.

3. Perinatal:

Prematurity or postmaturity.

Anoxia.

Prolonged, precipitate or complicated labour.

4. Postnatal:

Neonatal jaundice.

Convulsions.

Meningitis or encephalitis.

Congenital anomalies.

Middle ear disease, etc.

B. SYMPTOMATIC

These conditions present with signs or symptoms such as delays in milestones of head control, in use of hands, sitting, walking, attention to sound, babbling and speech, etc. When the mother suspects that all is not well, unfortunately she is often right and must be taken seriously.

Some agreed classification must eventually be formulated and adhered to, or confusion is inevitable; for instance, 'prematurity' is classified here as perinatal, but since its causes are also operating prenatally it is often included under the heading of 'prenatal' when causation is being investigated. Similarly, jaundice and its immediate causes could well be included under any of the above four headings. It is for these reasons that percentage figures for the relative importance of prenatal, perinatal, and postnatal causes of the conditions in question do not always reflect a comparable picture in different series.

The Role of the Medical Officer of Health

Finally, the administrative and practical arrangements necessary for the success of such a project will be discussed, and also the relative parts to be played by each section of our tripartite Health Service.

For the actual compilation and keeping of the Register, the Medical Officer of Health seems best place for the task. His department is familiar with, and already has considerable experience in, the organisation of such comprehensive programmes; it is also recognised by statute for the notification of births and infectious disease, and has statutory relationships with the Ministries of Health and Education, plus long-established arrangements with the Registrar-General. Also, its health area of practice is large enough to give significant figures, and the staff is of sufficient size and suitably qualified to collate and assess such data. Standardised and carefully drawn up forms would obviously be necessary to avoid the permanent loss of clinical information so vital for research purposes and as a pointer to the path ahead.

The majority of the types of cases listed above as suitable for a Risk Register do in fact eventually find their way on to the

handicap registers already kept by the Medical Officers of Health, who are also School Medical Officers. The Medical Officer of Health, under Section 22 of the National Health Service Act, 1946, already has a statutory duty to satisfy himself that adequate provision is made for the health and wellbeing of all children under 5 years of age in his health area, which includes the ever earlier diagnosis of the abnormal, and the scientific investigation and prevention and death and ill-health of all infants and young children. He has similar responsibilities for children over 2 years of age under the Education Act, 1944. The Ministries of Health and Education, in their circulars to Health Departments, both clearly show their anxiety that all handicapped or potentially handicapped children should be kept under review from birth onwards.

Further, those children on the Risk Register who do eventually prove to be handicapped are likely to suffer from unavoidable deprivations, from hospitalisations, and from many anxieties and frustrations associated with their handicaps. As a result of these, or through endogenous defects or other damage, they may suffer from special learning difficulties, even if their intelligence and other avenues of learning are apparently unaffected. The Medical Officer is thus intimately connected with the extremely difficult task of assessment and placement of these children in such situations as a normal school, a special class, or a special day or residential school, whichever is the most appropriate at the time. It is also his duty to forewarn the teachers so that kindly consideration can be given to the child, and the appropriate special teaching methods can be used for his or her benefit. A further important reason for choosing the Medical Officer of Health as the custodian of the Register is that, in contrast to the hospital paediatrician and the family

doctor, his area of practice possesses finite boundaries which can only be altered by law. Thus an address of a case must fall either in his area or in that of his neighbouring Medical Officer, and as a result administrative loopholes can be closed more easily.

Sources of Information

These sources vary, but the most important are:

1. The maternity services:
 - (a) The maternity hospital.
 - (b) The general practitioner and domiciliary midwife obstetric services.
2. The infant welfare services and health visitors.
3. The family doctor.
4. The hospital paediatric services.

Notifications of the discharge of mothers and babies from hospital are at present sent to the Medical Officer of Health, and arrangements could be made to include the 'risk' categories on such notifications; similar arrangements could be made with the midwives.

Besides the domiciliary midwifery and health visiting services already mentioned, the Medical Officer of Health also has under his administration other members of staff who are often both excellent sources of information for the Register, and subsequently, after diagnosis has been made, of great help in the support of the handicapped child and his family. These include assistant medical officers, home nurses, clinic and school nurses, audiometricians, orthoptists, physiotherapists, speech therapists, home helps and clerical staff. He is also in close, and often daily, contact with visiting teaching and Regional Hospital Board specialists, schoolteachers, peripatetic teachers, and social workers from the many voluntary societies for handicapped children. He also has in his files, going back many years, records of families who have had handicapped children, compiled at infant welfare clinics and by health visitors, and at a later stage school medical records.

Local Authority, Family Doctor and Specialist Panel

It is therefore proposed that the Medical Officer of Health, or a medical member of his staff, should be made responsible for the administration of a Risk Register, so that all relevant information from the various sources mentioned could without delay be channelled to him.

Our experience has shown that the Risk Register system will function only if it is fully accepted that the Medical Officer of Health is not only the custodian of the Register but also the person responsible for initiating the reference for assessment by various specialists and for follow-up procedures, and for the co-ordination of the child's treatment.

The Medical Officer of Health is then in a position to arrange the first diagnostic appointments, the timing of which is extremely important.

Before referral, every case must be discussed with the general practitioner, who must at all times be kept fully informed of the position. To obviate unnecessary anxiety on the part of the parents, the whole procedure must be fully and tactfully explained to them, but if this is done the modern parent will almost always be most appreciative of such comprehensive care.

The examination of functional systems, such as vision, hearing, or movements, in infants and young children is an exacting and complicated task demanding considerable experience, skill and insight; ability in these directions is a rare gift given to few. The choice of specialists, therefore, must take all these factors into account.

It is therefore suggested that the Local Health Authority (through its Medical Officer of Health), in conjunction with the Regional Hospital Board, should form in each health area a suitable panel of such specialists who must be willing to work as

a team, dedicated to the purpose of early diagnosis and where possible the prevention of disease.

The constitution of this panel of experts is beyond the scope of this paper, but in this Local Health Authority area we are fortunate in already having a cerebral palsy and ophthalmic unit attached to the local school for the physically handicapped, and an audiology unit attached to the school for the deaf. The visiting specialists are supplied by the Regional Hospital Board, and the rest of the medical and auxiliary staff, premises and equipment are administered and supplied by the Local Health and Education Authorities, under

Section 22 of the National Health Service Act, 1946, and the Education Act, 1944. All these units are in close proximity to one another, and also in close touch with the teachers and psychologists at the schools, resulting in a harmonious, rapid, and smoothly functioning system for the assessment, placement and treatment of the affected children, and the support of their families, in liaison with the family doctor.

In some other areas there are diagnostic and assessment clinics in existence in hospitals which co-operate with area Medical Officers of Health to give service for handicapped children.

SUMMARY

Too many children with defects are still being diagnosed too late for treatment to be fully effective.

The merits of three methods of detecting such defects, involving the central nervous, visual and aural systems, are discussed. The compilation of a Risk Register is suggested as an immediate and practical additional method to the present symptomatic method of detection, and a skeleton classification of aetiological categories is given.

The Medical Officer of Health is suggested as the custodian for the Risk Register, his area of practice being large, with fixed and finite boundaries, and his staff being already used to assessing and collating this type of information. For the system to be a success, he should also initiate the follow-up examinations and act as the team's co-ordinator.

The role of expert diagnostic teams, and their liaison with the family doctor, school-teachers and psychologists is discussed.

The primary purpose of a Risk Register is therefore to ensure earlier diagnosis during infancy, efficient and regular review, and appropriate management and treatment of those unfortunates who are born with a congenital defect.

RÉSUMÉ

Registre de risques

Il y a encore trop d'enfants déficients pour lesquels le diagnostic est fait si tardivement que le traitement ne peut plus être totalement efficace.

L'auteur discute des mérites de trois méthodes de détection de déficiences frappant le système nerveux central et les appareils visuels et auditifs. Il propose la constitution d'un 'registre de risques' qui pourrait compléter rapidement et de façon pratique la méthode de détection symptomatique actuelle et un squelette de classification de catégories étiologiques.

Il suggère que le médecin officier de santé soit chargé de ce 'registre de risques' entre autres parce que sa zone d'activité est vaste et a des limites bien déterminées et circonscrites. De plus, son équipe pourra toujours servir à récolter et confronter ce type de renseignements. Il faudrait pour que le système soit efficace qu'il soit aussi l'initiateur des examens ultérieurs et le coordinateur de l'équipe.

L'auteur discute du rôle d'une équipe de diagnostic expérimentée et de ses rapports avec le médecin de famille, les professeurs et les psychologues scolaires.

Le but essentiel d'un 'registre de risques' serait donc d'assurer un diagnostic plus précoce durant l'enfance, des contrôles efficaces et réguliers et un traitement et des soins appropriés à ceux d'entre nous qui ont eu la malchance de naître avec une déficience congénitale.

ZUSAMMENFASSUNG

Risiko Register

Zu viele Kinder mit Defekten werden noch zu Spät erkannt so dass die Behandlung nicht mehr völlig wirksam sein kann.

Die Vorzüge dreier Untersuchungsmethoden der Defekte, die das zentral-nervensystem und die seh und gehörsysteme befallen, werden besprochen. Die Einrichtung eines 'Risiko Registers' wird vorgeschlagen als unmittelbare und praktische Hilfsmethode und ein Klassifizierungs Entwurf der ätiologischen Kategorien wird gegeben.

Der ärztliche Gesundheitsoffizier wird als Hüter des 'Risiko Register' vorgeschlagen, weil unter anderen Gründen sein Wirkungsgebiet weit ist und feste Grenzen besitzt und sein Personal bereits gewöhnt ist, diese Art Auskünfte zu sammeln und zu vergleichen. Wenn dieser vorschlag angenommen wäre, müsste er damit sich dieses-system erfolgreich erweise, auch die folgenden Untersuchungen einleiten und das Personal koordinieren.

Die Rolle eines erfahrenen Personals für die Diagnose und die Beziehungen zwischen diesem Personal und den Familienarzt den Schullehrern und Psychologen wird erörtert. Das Hauptziel eines 'Risiko-Registers' ist folglich, eine frühere Diagnose im Kindesalter, wirksame und regelmässige Prüfungen und angemessene Pflege und Behandlung für diejenigen unter uns die das Unglück hatten, mit einem angeborenen Defekt zur Welt zu kommen.

REFERENCES

- Bordley, J. E. (1960) *In The Modern Educational Treatment of Deafness*. Ed.: A. W. G. Ewing. Manchester University Press.
- Drillien, C. M. (1958) 'Growth and development in a group of children of very low birth weight'. *Arch. Dis. Child.*, 33, 10-19.
- Fisch, L. (1955) 'Deafness in cerebral palsied schoolchildren.' *Lancet*, ii, 370.
- (1961) Personal communication.
- Gardiner, P. A., James, G. (1960) 'Association between maternal disease during pregnancy and myopia in the child.' *Brit. J. Ophthal.*, 44, 172.
- (1961) 'The development of myopia in early and later childhood.' *Cer. Pal. Bull.*, 3, 373-378.
- Harrison, K. (1960) *In The Modern Educational Treatment of Deafness*. Ed.: A. W. G. Ewing. Manchester: Manchester University Press.
- Howorth, I. E. (1958) 'Screening tests of hearing in pre-school children, with particular reference to selective testing.' *Med. Offr.*, 100, 307.
- Illingworth, R. S. (1960) *In 'Child neurology and cerebral palsy: early diagnosis.'* *Brit. med. J.*, ii, 1082.
- Mitchell, R. G. (1959) 'Medical aspects of a comprehensive survey of cerebral palsy.' *Cer. Pal. Bull.* 1, No. 7, pp. 32-41.
- Wellesley Cole, R. B. (1959) 'The problem of unilateral amblyopia. A preliminary study of 10,000 national health patients.' *Brit. med. J.*, i, 202.
- Woods, G. E. (1958) 'The early diagnosis of infantile cerebral palsy.' *Publ. Hlth.*, 2, 54.

The Dental Care of the Cerebral Palsied Child

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Successful treatment of the cerebral palsied depends on the co-operation of skilled individuals of different disciplines. This paper outlines the role of the dental surgeon in this team, and ways in which the other members can assist in the maintenance of healthy oral tissues in the cerebral palsied child.

A LARGE proportion of cerebral palsied children suffer from diseases and abnormal conditions of the mouth and teeth (Fig. 1). The incidence of dental caries, gingival disease and malocclusion is higher in this group than in the rest of the community. There are many reasons for this, since cerebral palsy can affect the oral structures directly and indirectly.



Fig. 1. Neglected mouth of an eleven-year-old boy, an appearance typical of that found in many cerebral palsied children.

Direct Effect of Cerebral Palsy

Because cerebral palsy is not a single condition but manifests itself in many ways, its effect on the oral tissues varies from one individual to another. In the predominantly athetoid child, involuntary movements of the face, teeth-grinding, abnormal tongue movements, and poor sucking, swallowing and chewing are common findings. Grinding the teeth produces flat occlusal

surfaces on the posterior teeth, eliminating to some extent the fissures in which decay often begins. However, because of difficulty in swallowing and inadequate oral cleansing due to poor muscle function, what advantage might have been gained from this wear is lost. Food particles lodge round and between the teeth and are soon broken down by oral bacteria, leading to caries and gingival disease.

Most of the findings in the predominantly athetoid are also found in the spastic and the ataxic child. However, abnormal tongue behaviour in these forms often results in an open bite in the incisor region (Fig. 2), which, together with the abnormal facial muscle patterns, provides the characteristic appearance of a narrow upper and lower arch, open mouth posture and protruding upper teeth. This appearance is not always found, for the eventual position of the teeth is determined by genetic as well as local factors. With the limited information available it would seem most unlikely that these individuals differ genetically from the rest of the ethnic group of which they are part.

Indirect Effects of Cerebral Palsy

The dietary habits of a child with cerebral palsy are governed both by the effect of the disease on the masticatory apparatus, and by the dietary habits in the home or institution. Because of the



Fig. 2. Open bite in the incisor region due to abnormal behaviour of the tongue muscles.

former, fibrous foods are rarely found in the diet, carbohydrates are largely taken in refined forms, and proteins and fat are taken mainly as milk. This in itself provides a reason for the unhealthy gingivae which are deprived of nature's tooth brush—hard, fibrous food. Furthermore, refined carbohydrates provide an ideal medium for the bacteria which produce caries.

Milk is, of course, beneficial, especially during tooth development, and later provides a cheap source of animal protein, fat, vitamins and minerals. Unfortunately, it is rarely given without an excessive amount of sugar. Dietary supplements in the form of sweets, cakes and biscuits are commonly given to cerebral palsied children, probably more than to normal children.

The tooth-brush normally provides a means of removing adherent food particles and stimulating the gingival tissues. The cerebral palsied child is often denied this procedure either because of his own inability to use it or because his parents find it difficult to brush his teeth for him.

Importance of Maintaining Oral Health

The main aim of the management of

children with cerebral palsy is to help them take their place in society. Dental care is obviously necessary for these children, for the social stigma of untreated or badly treated dental disease is very real, and an already handicapped child should not have an unnecessary additional handicap forced upon him.

Dental care is important aesthetically and functionally. Abnormal appearance resulting from the primary disease can be improved by good dental care, thereby helping the individual in his relationship with others. It is not only the child who benefits; the mother is very conscious of her child's appearance and welcomes any improvement that dentistry may give. She is as a rule very health-conscious, having been brought into close contact with the medical profession since the primary condition was diagnosed. To her, dentistry is just an additional service in the treatment of her child.

The teeth, though not essential for speech, are important for the production of clear sounds. Speech dysfunctions are common in cerebral palsy, and any oral and dental abnormalities, such as abnormal swallowing habits, poor muscle control and malocclusion, make the correction of speech defects more difficult. The improvement that the treatment of the oral conditions can bring is of undoubted value for these children.

The child who initially has difficulty in chewing and food management, may, with the aid of physiotherapy and speech therapy, eventually be able to use his oral mechanism quite well. However, unless the teeth have received adequate attention, full functional efficiency will never be obtained.

Dental care, then, is not the prerogative of the normal child alone; the handicapped child also requires a full dental service if he is to play his part in the community.

Treatment by the Dental Surgeon and its Attendant Problems

The dentist, confronted with the cerebral palsied child, must be aware of the nature of the child's handicap and able to apply



Fig. 3. Athetoid child receiving dental care. The operator is about to put a prop in place to prevent sudden closure of the mouth during treatment. The assistant is controlling spasmodic movement of the arms.

special skills and procedures to overcome it. He must be able to manage the random, uncontrolled movements of the athetoid child. Special technical skills include the use of gags to prevent injury to the surgeon's fingers and the soft tissues of the child's mouth (especially in the child who has exaggerated reflexes), the use of additional

help in controlling spasms, and the dental management of the child under general anaesthesia (Fig. 3).

Apart from the special technical skills which obviously require special training, he needs real compassion and understanding. He must appreciate the medical, mental and emotional problems that each child presents. He should understand the mother's role and her difficulties at home. His service must improve the child's dental health without further increasing the emotional burden on that child.

The difficulties in providing dental treatment for these children make prevention the primary objective. The dentist must be fully conversant with caries control techniques and should enlist the aid of other members of the team, especially the mother. Dietary control will, without doubt, reduce the incidence of decay. A simple rule is to reduce the intake of refined carbohydrate to the minimum, and confine it to meal times. In addition, tooth brushing should be taught either to the parent or the child, and be practised after every meal (Fig. 4).

In Great Britain, the dental services for children are already overburdened, primarily because there is a lack of qualified dentists. The training of ancillaries who will treat children's teeth has begun, but it is debatable whether these people should ever undertake the management of the physically and mentally handicapped child. What is required is a special service, staffed by fully trained personnel. This might come within the National Health Service or be separate from it. Specially designed clinics with full facilities are necessary if dental care for handicapped children is to be a reality.



Fig. 4. The teeth of an athetoid child being brushed by her mother. The position has been specially chosen as being one in which there is the least abnormal movement of the head or limbs.

SUMMARY

The maintenance of healthy oral tissues in cerebral palsied children is not only the responsibility of the dental profession. Co-operation is needed in this work.

Dental disease is to a great extent preventable. Refined carbohydrates, such as sweets and other confections, are largely to blame for dental caries, and in the cerebral palsied child they should be reduced to the minimum and only taken at meal-times. Any remaining food must be removed by tooth brushing, which should be taught to both the child and his parent, and practised after every meal.

Specially designed clinics are needed if dental care for these children is to be a reality.

RÉSUMÉ

Les soins dentaires de l'enfant encéphalopathe

Les membres des professions dentaires ne sont pas seuls responsables du maintien en bon état de santé des tissus buccaux des enfants encéphalopathes. La coopération est nécessaire en cette matière.

On peut prévenir les affections dentaires dans une large mesure. Les hydrates de carbone raffinés, tels que les bonbons et autres préparations, sont, dans une large mesure, responsables des caries dentaires, et chez les enfants encéphalopathes il y a lieu de les réduire au minimum et de ne les absorber qu'aux heures des repas. Il faut apprendre à l'enfant et à ses parents que les aliments résiduels doivent être éliminés en brossant les dents et que ce brossage doit être pratiqué après chaque repas.

Des cliniques spécialisées sont nécessaires si ces enfants ont besoin de soins dentaires.

ZUSAMMENFASSUNG

Die Zahnpflege beim Kind mit Zerebrallähmung

Für das Gesundhalten der Gewebe des Mundes bei Kindern mit Zerebrallähmung ist der zahnärztliche Beruf nicht allein verantwortlich. Zusammenarbeit ist für diese Arbeit nötig.

Zahnkrankheiten sind in grossem Masse verhütbar. Raffinierte Kohlenhydrate wie Bonbons und anderes Konfekt sind zum grossen Teil an Zahnkaries schuldig und sie müssen beim Kind mit Zerebrallähmung aufs Minimum herabgesetzt und nur während der Mahlzeiten genossen werden. Man muss dem Kinde und seinen Eltern beibringen dass alle Speisereste durch bürsten der Zähne, nach allen Mahlzeiten zu beseitigen.

Spezialisierte Kliniken sind nötig wenn Zahnpflege für diese Kinder eine Wirklichkeit sein soll.

NOTICE

National Bureau for Co-operation in Child Care

THE National Council of Social Service has now set up an organising committee, charged with the task of bringing into being the National Bureau for Co-operation in Child Care, which recently held its first meeting. One of the first problems to which the Committee is giving its attention is the securing of the necessary finance to establish the Bureau on a sound footing and to help it to develop.

The proposal to set up such a Bureau was first mooted almost ten years ago at a meeting of the Association of Children's Officers, and discussions and negotiations have been proceeding in various quarters ever since that time. The National Council of Social Service was brought into the picture in 1959 and, in that and the following year, organised three conferences of people concerned with child care in all its aspects. At the last of these conferences, held at Church House, Westminster, in December 1960, in the presence of some 130 representatives of 80 voluntary organisations, professional associations, universities and Government and local authorities, the proposal received wholehearted acceptance and it was agreed that the N.C.S.S. be asked to take the necessary action to bring into being a Bureau of Child Care, whose broad aims would be to improve communications between workers (voluntary and statutory) in the various child-care fields and to provide a service of information and advice.

One of the Bureau's initial activities will be the collection of information about research and the establishment of a reference centre where relevant information about the work done in this country and abroad could be indexed and kept up-to-date. Close contacts will be developed with universities, professional associations (social, educational and medical); new projects for research and enquiry will be studied and means of consultation established with the appropriate bodies for the furtherance of relevant enquiries. An integral part of the Bureau's activities will be the establishment of regional and local groups of professional and lay people actively engaged in various child-care services. It will provide guidance on the methods and organisation of these local groups and supply them with information, and in return receive suggestions and views on practical problems which require study. It will arrange conferences and seminars to provide opportunities for individuals working in different services to meet together for joint study and discussion of common problems.

Membership of the Bureau will be open to authorities, organisations and individuals. It is proposed that it shall be centred in London, easy of access, and able to call on the best knowledge and skill available.

WHAT'S TO BE DONE?

Problem:

Are children with athetoid cerebral palsy nearly always thin? If so, why? Is a metabolic factor involved? What special steps can be taken to correct this tendency, from either the nutritional or the pharmaceutical angle?

Replies:

1. *From the Staff of the Dame Hannah Rogers School, Ivybridge, Devon.*

We agree that most children with athetosis are thin, though we have not got a large enough number of children under review at this school to provide a statistically significant answer. One definite factor is the constant activity of these children, and the considerable amount of energy they use up by their excessive muscular contractions. We are not aware of any work that has confirmed the existence of any additional metabolic factor.

In our opinion it is desirable that these children should be rather on the thin side, from the point of view of their mobility, and we are not usually anxious to increase their weight to any great degree. In general terms, our preoccupation at this school is to prevent our children becoming overweight, rather than the reverse.

2. *From Prof. J. CIZKOVA, University Children's Hospital, Prague, Czechoslovakia.*

It is well known that the majority of patients with cerebral palsy tend to get too fat, but that the athetoid children are usually thin. Here are some possible explanations:

- (1) There may be distortion of the C.N.S. centres which regulate the appetite and thereby the intake of food.
- (2) The centres which regulate the enzymatic system of the gastrointestinal tract may also be affected.
- (3) Poor mastication.
- (4) Incorrect swallowing.
- (5) Neglected home care.
- (6) The muscular hyperkinesia of these children takes up a great deal of energy, and in athetoids the intake of food is not economically distributed.

REPORTS

Evaluation of Treatment in Cerebral Palsy

Seminar organised by the World Commission on Cerebral Palsy

Vienna: August 18-20, 1961

Reported by Dr. P. HUME KENDALL

Guy's Hospital, London, S.E.1.

Objectives

THE purposes of this informal gathering, to which 35 participants came from the United States, England and Scotland, France, Belgium, Italy, Germany, Austria, Denmark, Norway, Sweden, Finland, Brazil, South Africa, and Australia, were (1) to share experiences in the treatment and management of the various types of cerebral palsy; (2) to ascertain the feasibility of agreement on an objective evaluation of the effectiveness of various modalities of total treatment; and (3) to explore the possibilities of collaborative efforts in evaluating current treatment methods. The meeting opened with greetings from **Dr. Guy Tardieu**, Chairman of the World Commission, and **Dr. Donald V. Wilson**, Secretary of the International Society for the Rehabilitation of the Disabled. **Dr. Brewster S. Miller**, Secretary of the World Commission, who was responsible for the arrangements, explained the seminar's aims and programme.

British, American and French Views on Assessment

Dr. P. Hume Kendall (*Guy's Hospital, London*) gave a report on recent joint discussion on assessment between the N.S.S. and British Council for the Welfare of Spastics. The meeting was largely concerned with the difficulties of achieving a uniform method of assessment of such a diverse problem. Most speakers were doubtful about even commencing a pro-

ject but several, including Prof. A. A. Moncrieff, Prof. R. S. Illingworth, Dr. Ellis and Dr. Hume Kendall, thought it was essential to attempt to find a solution. **Dr. Kendall** now felt that that meeting had been a failure in as much as there were no concrete suggestions. He made a plea for agreement, if possible, on the various types of proforma required to record the various aspects of the patient's disability and the development of a uniform protocol. Once established the World Commission should co-ordinate a trial of physical therapy or certain aspects of it for a period of, say, three years.

Dr. Raymond R. Rembolt (*Iowa*) described the American Academy of Cerebral Palsy's project to develop an assessment scheme for the objective evaluation of the therapy of cerebral palsy. A committee had been set up which deliberated on the problem for two years before arriving at what he described as a 'concrete solution'. They had developed a plan to assess the whole child as far as possible. This was to be accomplished through two groups: (a) a team going out to various centres to evaluate suitable children twice yearly; and (b) a central evaluation team of statisticians. The scheme would be confined strictly to patients who conformed to the present diagnostic criteria of cerebral palsy. Initially the team would be concerned only with untreated patients and could not be confined (at first at least) to any age-group, intelligence level or degree of motor disability. Once the child was assessed a

therapy regime would be prescribed and subsequently followed up. Various centres would be encouraged to make use of the visiting team. The team would take a special interest in home therapy, as well as therapy in a centre, in areas in which no therapy was available or where only very limited therapy could be obtained. The team would consist of:

1. Paediatric neurologist.
2. Physical medicine specialist or orthopaedist.
3. Psychologist.
4. Secretary.
5. Photographer.

For the first six months this team would conduct a pilot trial to get experience with methods of recording. They would be based on a university setting and would eventually aim to conduct a survey for not less than 5 years and probably up to 10.

Dr. Margaret H. Jones (*Los Angeles*) described the proposed assessment charts of the American Academy of Cerebral Palsy project. These consisted of:

- | | |
|---|---------------------|
| (a) A summary sheet. | |
| (b) A graph of general progress. | |
| (c) An inclusive history and examination sheet for the paediatrician. | |
| (d) Neurological | } assessment charts |
| (e) Orthopaedic | |
| (f) Speech | |
| (g) Otological | |
| (h) Psychiatric | |

The psychiatric assessment would include social and educational evaluation and various tests of intelligence. X-ray pictures of the pelvis would be taken and the child photographed and filmed. An effort was being made to keep the assessment charts reproducible and intelligible, and in a form suitable for statistical survey.

There was then considerable discussion about controls for the various forms of therapy. It was eventually agreed that it would be possible to compare treatment A with treatment B, thereby avoiding the

necessity for completely withholding therapy which raised ethical problems. **Prof. R. S. Illingworth** (*Sheffield*) said this difficulty could also be overcome by alternating periods of treatment and no treatment.

Mr. George Pollock (*Edinburgh*) raised the point that allowance would have to be made for improvement due to natural maturation which should not be attributed to therapy.

Dr. Adriano Milani-Comparetti (*Florence*) made a plea that this form of assessment should be confined to research and not used to decide whether a child should be admitted or rejected from a centre for therapy.

Dr. Guy Tardieu (*Paris*) then outlined his work with the electromyograph and strain gauges. He had detected that there was a certain speed limit at which a spastic muscle could be stretched without exciting electrical activity. He also described his method of computing electromyographic potentials into quantitative units by means of an electrical integrator. He showed that there was a constant relationship between the quantity of electrical activity and the rate at which a muscle was stretched. He thought it was possible to use these methods to assess objectively and they might be used to test the action of drugs or physical therapy on spasticity.

The Treatment of Hemiplegia

Dr. C. Burton-Bradley (*Australia*) described her routine management of the spastic hemiplegic patient in her centre in Sydney. She had analysed 134 cases out of a total of 1,000 with cerebral palsy and preferred conservative management, resorting to surgery only as a last resort.

This provoked a discussion on the advisability or otherwise of early surgery. There was clearly a wide difference of opinion here. It seemed generally agreed, however, that triple arthrodesis of the ankle, rotational osteotomy of the knee-

joint, and adductor tenotomy and arthrodesis of the wrist-joint were the most successful forms of surgery. There was agreement that tendon transplants were not generally successful. It was also agreed that an effective postoperative regime of therapy was essential for good results.

Dr. William A. Hawke (*Canada*) mentioned the high incidence of behavioural difficulties and sensory loss in hemiplegics. He felt that there was no point in treating them before school age but observed that the older they became the more difficult they were to treat. In discussing behavioural problems, many speakers pointed out that epilepsy was more common and psychological disturbances frequent in the teen-aged hemiplegic.

There was much disagreement on the place, type and timing of physical therapy in the hemiplegic. **Dr. Karel Bobath** (*London*) felt that it was useful, particularly where there was a sensory disorder; early treatment was then vitally important.

Dr. Ben Epstein (*South Africa*) wondered whether spastic hemiplegia might not prove to be a useful starting point for any scheme of assessment. Dr. Kendall agreed, pointing out that there were quite a few definite problems which could be answered by accurate assessment and follow-up.

Dr. David D. Rutstein (*U.S.A.*) then observed that so far the meeting had not attempted to evaluate therapy but each speaker had said what he felt was the best form of management for hemiplegia. Speaking as a statistician, he pointed out certain fallacies with regard to the statistical analysis of patients selected or rejected for a particular form of therapy but assured the meeting that there were various approved methods of handling sampling that were suitable for such an evaluation; these should be used.

Dr. Preston Robb (*Canada*) suggested that it was time to review the main aims of the seminar.

Dr. J. P. Duarte (*Brazil*) then outlined his system of assessment into 5 grades according to the child's mental age. This was divided into 0-4 months, 4 months-2 years, 4-6 years and 6-7 years. For each group a routine of therapy had been developed.

Dr. Marcel D'Avignon (*Sweden*) emphasised the need to evaluate a patient by joint consultation with a team of physicians and surgeons after a period in which all had had time to know the child. It was important to bring nurses, therapists and teachers into this discussion. Several other speakers knew of patients who had received intensive comprehensive physical therapy for many years without improvement. They wondered whether in fact therapy was justified in these types of cases and agreed that objective evaluation was required.

Dr. Guy Tardieu said one must be careful not to do too much but also not to do too little. **Professor Illingworth** said that now was the time to study a list of the various forms of evaluation of therapy in use.

Dr. Rutstein then discussed at length the planning of an investigation of therapy in cerebral palsy. First, there were two main questions to consider:

(1) Was one going to take a group of patients and study them thoroughly, making records of every possible feature?

Or (2) was one going to attempt to investigate a single question?

The first approach, though the most thorough, was lengthy and time-consuming. It would yield much information on the natural history of cerebral palsy but was unlikely to answer specific problems.

The second method would provide an answer only to the question asked. Any further problem would mean starting from the beginning and planning the trial over again. If this second type of trial was

envisaged, there were several important problems to be decided beforehand:

1. Was the problem an important one?
2. Was it feasible to solve it?
3. What kind of patients with cerebral palsy were to be selected to answer this problem?
4. What kind of information (or measurements) were needed to produce a result?
5. What would be the criteria of success or failure of the trial? For instance, would one judge the effect of an orthopaedic procedure purely so far as it affected limb function or would one consider how it eventually affected the whole child?
6. What data is to be collected, how was it to be collected, and how was it to be analysed? The means of analysis—computers, punch-cards, etc.—must be made to fit the requirements, rather than the requirements being made to fit the machines.

When selecting the patients it was important to have accurate definitions to ensure that everyone selected in the same way. Therefore, the next problem to decide was what were the most favourable patients, problems and techniques for investigation.

The Evaluation of Treatment of Athetosis

Dr. Robb felt that physical therapy had little or no part to play in the treatment of athetoid cerebral palsy. This resulted in a great many speakers delivering their view, but largely the feeling was that physical therapy had little or no active part to play. Nevertheless, no one was certain of this, and they therefore continued to prescribe physical therapy because there was little else to be done. **Dr. J. Siebert (Germany)**, however, went so far as to say that he never used physical therapy for athetoids and they got on just as well.

Dr. Robb said it seemed generally agreed

that physiotherapy did little to reduce the disability that resulted from athetosis, but he wondered if we could formulate methods of assessing the disease and its therapy. **Professor Illingworth** again pointed out we would not only have to assess the patient at any one time but also allow for the improvement associated with maturation.

Dr. Tardieu felt that assessment could be considered in two ways:

- (a) Overall assessment of the patient which could be done well with a suitable film.
- (b) Local assessment; testing of particular relevant facets of the motor disability.

Each participant was then asked to state his present methods of objective assessment. These were given only generally, without details. **Dr. Tardieu** used both electrodiagnostic and clinical evaluation. **Dr. Bobath** claimed he had developed an accurate method of functional tests. **Dr. B. Anderson (Norway)** used a combination of the McGraw tests and reports from the therapists. **Professor Illingworth** mentioned simple tests that did not rely on much apparatus, such as walking, footprints, dialling, screwing, typing, pressing a bell, etc., all of which could be timed.

The American Academy of Cerebral Palsy tests were briefly surveyed and the 5-point scales in use at Guy's Hospital were mentioned. The Gesell, Ozaretsky and Albitreccia scales were also widely used.

Several participants pointed out the importance of recording the conditions at the time of the test, and the need for uniformity; and for relating the results to the patient's age. The posture of the patient when performing the test should also be recorded and visual acuity and hearing tested. The child's motivation for the tests considered. The tests must be pleasant and interesting. Two other features were agreed to be necessary—first, space for impressions to be recorded, and secondly

an attempt at a prognosis at the end of the evaluation.

Drug Therapy

In the discussion on drug therapy in choreo-athetoid cerebral palsy there was a large measure of agreement that the results were generally disappointing. Some participants thought there was a place for selected drugs. **Dr. Tardieu** and **Dr. Kendall** used meprobamate and carisprodol (Soma) in certain patients. **Dr. Milani** found primidone (Mysoline) and carisprodol (Soma) or a combination of the two, of value in severe dystonia. **Dr. Epstein** and **Dr. Bobath** found the tranquillizing effect of thioridazine (Melleril) of value in patients with mental defect. **Dr. Jones** had seen two patients with athetosis treated with chlordiazepoxide (Librium) with good effect, and it was reported that Professor Plum favoured this drug. On the other hand, when asked by **Professor Illingworth** whether these were the results of clinical impressions or double-blind controlled trials all the speakers admitted they were relying on impressions only, though **Dr. Tardieu** argued that this did not matter. **Professor Illingworth** reported that in three double-blind trials of primidone, trihexyphenidyl (Artane) and carisprodol he had shown that there was no significant difference between the activity of these drugs (in doses recommended by the manufacturers) and placebos. **Dr. Rembolt** and others reported the same findings in similar trials.

Dr. Karel Bobath showed two films, one on the treatment of athetoid cerebral palsy and the other on the management of a child with cerebral spastic diplegia who had been committed to a mental institution as a mental defective and was suffering largely from emotional and sensory deprivation.

Methods of Recording Progress

Dr. William Hawke presided over a

meeting which was held to try to reach agreement on methods of recording the effectiveness of therapy in various aspects of cerebral palsy. **Dr. William Cooper (U.S.A.)** gave an account of the assessment system he used at the Hospital for Special Surgery, New York. This comprises two main groups: firstly a series of clinical history and examination forms which include a summary sheet, comments on prognosis, educational prospects, social and emotional problems, school placement and a programme design. Parallel with this is a series of tabulation charts on which numerical coding is allotted to 31 aspects of the history and examination, covering a wide range of features such as type of involvement, hereditary factors, current accomplishment levels, estimated prognosis, etc. The numeral allotted to each grade is tabulated on a chart and also recorded on a punch-card. The punch-card therefore provides a comprehensive record of the patient and previous therapy which accompanies the child, thus providing accurate information for everyone. There was general approval of this type of assessment though some disagreement as to the details in various areas.

There was then a lengthy discussion on how much information should be given to parents and at what stage in the child's development the full significance of its condition should be divulged. It was generally agreed that this information should be given at the earliest possible moment, but that the diagnosis should be withheld if there was any uncertainty, particularly if mental retardation was involved. Many agreed with **Dr. Chester A. Swinyard (U.S.A.)** who thought that information should be given to the parents as an educational process over a long period. **Dr. Donald Wilson** had always found absolute honesty essential, whether in giving a diagnosis or in divulging the degree of disability or the likelihood of

response to therapy. This same principle applied when making an appeal for funds; we should never say we were aiming to do what we could not do.

Management of the Cerebral Palsied Child

In discussing socialisation of the cerebral palsied child, **Dr. Wilson** felt that much of the progress of these patients depended on the local community. If there was suitable employment available on a sufficient scale they would get it; if not, they would not. **Dr. Margaret Jones** disagreed, saying that the small percentage of cerebral palsied patients who were going to obtain employment would do so whatever the circumstances and 90 per cent of the others would not—despite a very favourable environment. The difficulty in fitting children into normal social surroundings was discussed at length. There was considerable argument as to whether it was more desirable to establish special schools for cerebral palsied children or to arrange for them to receive adequate attention and education at normal schools, provided the degree of handicap was not great. It was felt that this was very much a social problem which probably varied in every country. **Professor Illingworth** was worried about adding an educational handicap to the physical handicap if one sent a child to a special school, because the level of education could never be as high as at a normal school and there was always a great expenditure of time on the various forms of medical therapy. However, it must be recognised that if a handicapped child was to maintain an average standard at a normal school it must be of above average intelligence.

Dr. Preston Robb described the special social discussion groups held by the older children at his special school, in which normal children participated. **Dr. Kendall** agreed that this was of great value. At the Thomas Delarue School in Tonbridge,

Kent, the children of school-leaving age were also sent on an independence week, when they did everything for themselves without help. This was of great use in building up morale and adjustment but was of inestimable value from the point of view of assessment of the difficulties that were to be expected in daily life.

Speech Therapy

It was agreed unanimously that speech therapy played an essential part in the programme of a cerebral palsy unit but there seemed to be considerable difficulty in its evaluation. The prognosis with regard to speech depended on the degree of involvement but there did not seem to be any way of determining which patient should receive specific speech therapy. Tape-recording with play back to a selected audience of observers seemed to be the most favoured technique for the clinical assessment of speech. **Dr. Tardieu** described his method of clinical assessment of speech but pointed out that the results might be different for other languages than French. **Dr. Swinyard** agreed; languages differed in the number of phonemes used, some Pacific tribes having as few as 13 whereas the average was around 30. He wondered if it might not be easier to teach those that use the fewest phonemes. There were differences of opinion on the exact meaning of 'aphasia', **Dr. Tardieu** in particular mentioning that he did not use the term. **Dr. Milani-Comparetti** felt that the important point to assess was not whether the child could speak but whether he could make use of the speech.

Psychotherapy

Dr. Donald Wilson said that people took for granted that psychotherapy by counselling was helpful but he wondered whether anyone had tried to assess its value. Much counselling was haphazard and probably useless. **Dr. Sven Brandt**

(Denmark) would make more use of tape-recording for the assessment of counselling. Several speakers, including **Dr. Hawke**, **Dr. Rutstein** and **Professor Illingworth**, were sceptical of the value of psychotherapy. **Dr. Duarte** suggested that very often it was not the child that required psychological assistance but the parents. **Dr. Ben Epstein** raised the important point that the child must be exposed to the correct stimuli if it was to develop its personality to the maximum; he therefore felt that schooling should be as normal as possible. **Dr. K. Bobath** thought the normal child looked for its own stimuli but the cerebral palsied child had to have its stimuli taken to it. **Professor Illingworth** agreed that one could retard a child by depriving it of normal stimuli, but one could not accelerate maturation beyond a certain rate however much one stimulated the child.

In a brief discussion on perceptual loss, **Dr. J. B. M. Hariga** (France) described the technique for assessing the 'gnosique' age, as practised in Dr. Tardieu's clinic. Various geometric forms were given to the patient to handle behind a screen. The results were correlated with a known scale and the resultant 'gnosique age' was compared with mental and chronological age. They had not found any relationship between defects of 'gnosique' appreciation and limb length but had noted that spatial appreciation could be improved by repeated handling of geometric figures.

Hydrotherapy

Dr. Lea Ylppo (Helsinki) described the Finnish sauna baths, which most of their children were given. She felt that they were of little more than psychological value but since almost everyone in Finland took them, the cerebral palsied child must do

so too. The resultant hyperpyrexia might precipitate a seizure in an epileptic child. **Dr. Kendall** thought that swimming was a useful competitive recreation in which the handicapped child could take part, but that hydrotherapy had no specific therapeutic effect.

Functions of the World Commission

Dr. Preston Robb put forward his personal view of the functions of the World Commission:

1. To help countries in developing their own services for the cerebral palsied.
2. To try and work out a uniform classification and language for describing cerebral palsy.
3. To make available the various evaluation systems, proformas and methods of assessment that had been described at the seminar, the Commission to act as a central agency for their distribution.

Conclusion

Professor Illingworth, in closing the Seminar, said it had been extremely valuable in teaching the participants to develop a healthy scepticism about all forms of therapy, drug or physical. No one would lightly accept the value of any treatment in future unless it was backed by suitable controlled trials. We must be sceptics but not therapeutic nihilists; there was a big difference between saying that a treatment was of no value and saying we did not know whether it was of any value. Above all, one must be sure that treatment was not harmful.

Two films were then shown, one from Poland on the Management of the Cerebral Palsied Child, and the second a beautifully produced sound-colour documentary from Canada, aimed at helping parents.

Schizophrenic Syndrome in Childhood

Progress Report of a Working Party (April, 1961)

DURING 1958, with the help of the Mental Health Research Fund, a re-examination was made at the Hospital for Sick Children, Great Ormond Street, of the long-term records of a group of severely disturbed children, diagnosed as psychotic. A number of these children had also been seen by other workers, and in the course of time had been variously classified as 'autistic', 'schizophrenic', 'atypical', sometimes with an additional or alternative label 'brain-damaged', 'epileptic', 'mentally defective', and so forth.

Diagnostic classification of schizophrenic illnesses in childhood rests ultimately on a clear understanding of the causes. Present knowledge, however, is inadequate for an attempt to be made to draw up a classificatory scheme on such a basis. Kallmann and Roth (1956) consider that in at least some pre-adolescent schizophrenics there may be an early effect of the same genotype assumed to be responsible for the basic symptoms of adult schizophrenia. The varied terminology—autistic (Kanner 1943), symbiotic (Mahler 1952), schizophrenic (Bender 1958) and others—makes it particularly difficult to know the extent of the clinical field covered by these terms. Bender's definition is perhaps the widest and most all-embracing, and she, perhaps more than others, sees this as a not infrequent occurrence in childhood. A brief list of references is appended; a comprehensive one is given in the section on *Childhood Schizophrenia and Allied Conditions* in Bellak's 'Schizophrenia', published in 1958.

In this country some clinicians concerned with the diagnosis of such children hold the view that the condition is less

rare than had been thought, and that more such children are being referred for psychiatric assessment. Similar cases are by no means unknown in hospitals for the mentally subnormal; but we could discover no generally applicable criteria which led on the one hand to referral to a child guidance clinic or hospital department of psychological medicine, or, on the other, to ascertainment as severely subnormal and referral to M.D. hospital or training centre.

With this situation, it was impossible to gain any idea of the extent of the problem. A few child guidance clinics were approached, but it was hardly surprising to find that the ratio of such children to the rest of their referrals varied considerably from clinic to clinic. The criteria used by the referring doctor, or the particular psychiatric viewpoint adopted, may be two of many reasons for such variations.

One result of these earlier studies was a meeting in May 1960 (by invitation of the research committee and the Board of Governors of The Hospital for Sick Children) of a group of 13 interested workers drawn from the fields of child psychiatry, paediatrics, genetics, psychology, psychiatric social work and mental deficiency. Obviously many other participants could have been invited, but it was felt to be of overriding importance to keep the group small enough to work intimately on a basis of discussion.

The agenda for this meeting proposed:

- (1) To discuss the present confused terminology and perhaps reach some clearer definition of the different

clinical syndromes and agree on the terminology.

- (2) To discuss the possibility of agreeing on a simple form which could then be circulated to clinicians who are likely to meet with these cases.
- (3) To discuss how best to use this material.

In all, six meetings (mostly of two sessions' duration) have been held between May 1960 and February 1961.

It was agreed that the first necessity was to clarify and describe what we meant by the term 'psychosis in childhood', and in this communication we offer the results of our attempts to do this.

Some members of the working party preferred the more general term 'psychosis in childhood' and others the more specific 'childhood schizophrenia'. After lengthy discussion it was agreed that the term 'schizophrenic syndrome in childhood' should be used. Nine diagnostic points were eventually generally accepted. We at first attempted to confine ourselves to a description of clinical signs and of behaviour which could be directly observed in interview. This proved to be impossible if we were to convey what we all felt to be the heart of the matter—namely, the presence of an impaired capacity for human relationships, which observation alone, however acute, cannot discover. Some aspects of these 'points', therefore, inevitably embody interpretations of behaviour, whilst others are more likely to be obtainable only from the child's history.

These nine points were not intended as absolute criteria in the sense that all, or any particular one, must be present; nor were they designed for use as a rating scale. While no attempt was made to arrange them in order of importance, it was found when they were 'tried out' by different members of the group (on four samples totalling 68 children who had

already been diagnosed as psychotic or schizophrenic) that the first point was present in every case except one. Many of us regarded the ninth point also as a *sine qua non*.

The Nine Points

1. Gross and sustained *impairment of emotional relationships* with people. This includes the more usual aloofness and the empty clinging (so-called symbiosis); also abnormal behaviour towards other people as persons, such as using them, or parts of them, impersonally. Difficulty in mixing and playing with other children is often outstanding and long-lasting.

2. *Apparent unawareness of his own personal identity* to a degree inappropriate to his age. This may be seen in abnormal behaviour towards himself such as posturing or exploration and scrutiny of parts of his body. Repeated self-directed aggression, sometimes resulting in actual damage, may be another aspect of his lack of integration (see also point 5), as also the confusion of personal pronouns (see point 7).

3. *Pathological preoccupation with particular objects* or certain characteristics of them, without regard to their accepted functions.

4. *Sustained resistance to change in the environment* and a striving to maintain or restore sameness. In some instances behaviour appears to aim at producing a state of perceptual monotony.

5. *Abnormal perceptual experience* (in the absence of discernible organic abnormality), implied by excessive, diminished, or unpredictable response to sensory stimuli—for example, visual and auditory avoidance (see also points 2 and 4), or insensitivity to pain and temperature.

6. Acute, excessive and seemingly illogical *anxiety*. This is a frequent phenomenon and tends to be precipitated by change, whether in material environ-

ment or in routine, as well as by temporary interruption of a symbiotic attachment to persons or things (compare points 3 and 4, and also 1 and 2). (Apparently commonplace phenomena or objects seem to become invested with terrifying qualities. On the other hand, an appropriate sense of fear in the face of real danger may be lacking.)

7. *Speech* may have been lost, or never acquired, or may have failed to develop beyond a level appropriate to an earlier stage. There may be confusion of personal pronouns (see point 2), echolalia or other mannerisms of use and diction. Though words or phrases may be uttered, they may convey no sense of ordinary communication.

8. *Distortion in motility patterns*—e.g., (a) excess as in hyperkinesia; (b) immobility as in katatonia; (c) bizarre postures, or ritualistic mannerisms, such as rocking and spinning (themselves or objects).

9. *A background of serious retardation* in which islets of normal, near normal or

exceptional intellectual function or skill may appear.

We do not doubt that further improvements could be made, and it is with this aim in view that we offer our contribution at this stage for comment and criticism—based preferably on usage.

Conclusion

A general agreement among psychiatrists on such diagnostic features would clear the way towards a common understanding and recognition of the phenomenological composition of the syndrome. A more immediate hope is that this attempt to extract from the observed data those signs of diagnostic value which are most frequently seen may lead to early recognition by those who are in a position to observe deviations in the development of infants and young children.

Offers of collaboration (forms would be supplied), or other communications arising from this, will be welcomed by the chairman of the group.

Members of the Working Party

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 DR. VALERIE COWIE, *Psychiatric Genetics Research Unit, Medical Research Council.*
 MISS SYLVIA INI, *P.S.W., formerly in juvenile psychosis research project at the Hospital for Sick Children, Great Ormond St., London.*
 DR. RONALD MAC KEITH, *Children's Department, Guy's Hospital, London.*
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REFERENCES

- Bellak, L. (1958) 'Childhood schizophrenia and allied conditions.' *In Schizophrenia*. New York: Logos Press.
 Bender, L. (1958) 'Psychiatric problems of childhood.' *Med. Clin. N. Amer.* **42**, 755.
 Despert, J. L., Sherwin, A. C. (1958) 'Further examination of diagnostic criteria in schizophrenic illness and psychoses of infancy and early childhood.' *Amer. J. Psychiat.*, **114**, 784.
 Eisenberg, L. (1957) 'The course of childhood schizophrenia.' *Arch. Neurol.*, **78**, 69.
 Hirschberg, J. C., Bryant, K. N. (1954) 'Problems in the differential diagnosis of childhood schizophrenia.' *Ass. Res. nerv. ment. Dis. Proc.*, **34**, 454.
 Kallmann, F. J., Roth, B. (1956) 'Genetic aspects of pre-adolescent schizophrenia.' *Amer. J. Psych.*, **112**, 599.
 Kanner, L. (1943) 'Autistic disturbances of affective contact.' *Nerv. Child.*, **2**, 217.
 Mahler, M. S. (1952) 'On child psychosis and schizophrenia.' *Psychoanal. Study Child.*, **7**, 286.

- Norman, E. (1954) 'Reality relationship of schizophrenic children.' *Brit. J. med. Psych.*, 27, 126.
 Putnam, M. (1955) 'Some observations on psychosis in early childhood.' In *Emotional Problems of Early Childhood*. Ed. G. Caplan. London: Tavistock Publications, p. 503.
 Strohm, G. (1960) 'On the diagnosis of childhood psychosis.' *J. Child Psychol. Psychiat.*, 1, 238.
 Symposium on Childhood Schizophrenia, 1955. (1956). *Amer. J. Orthopsych.*, 26, 499.
 Winnicott, D. W. (1953) 'Psychoses and child care.' *Brit. J. med. Psychol.*, 26, 68.

NOTICE

Cerebral Palsy in France

FRANCE has her own National Spastics Society, and its organisers would like this to be more widely known. Formed in 1956, L'Association des Infirmes Moteurs Cérébraux is divided into regional branches and is supported by public contributions and by Government and local authority grants. Its members include the families and parents of affected children, the medical and therapeutic staff of schools and homes for the cerebral palsied, and others interested in their welfare.

One of the Association's main objects is to increase public awareness of the cerebral palsy problem and to enlist public support and understanding. It hopes also to promote co-operation between other national and international organisations with similar interests. For this it publishes an informative bulletin, which compares with the *Muscular Dystrophy Newsletter* rather than with the *Cerebral Palsy Bulletin*. It is also engaged on an encyclopaedic work dealing with the medical aspects of cerebral palsy, which is to be produced in serial form. The first of the '*Feuillets*' has already appeared.

The Association's achievements, in the social, educational and medical fields, are already impressive. Among its other enterprises intended to help the affected children's families as well as the children themselves, are summer holiday camps, which are proving highly popular. In December, 1959, with Government support, the Association organised its first pilot Pre-School Centre, where 40 children aged 3 to 6 years are housed in a building specially designed and equipped to meet their needs. There is a kindergarten and wards for specialised and therapeutic education. The running costs are borne by French Social Security, and the centre is under the medical supervision of Dr. Graveleau. Most of the staff have been trained in the U.S.A. or in Great Britain, thanks to financial assistance from the United Nations European office and the French Ministry of Public Health. The centre is intended to be the precursor of similar schools for older children at secondary and technical levels, and it is hoped to build technical workshops and scientific laboratories for their use in due course.

The voluntary advisory medical committee which has been formed on behalf of the Association is an impressive body of 70 specialists from all over France, with the object *d'entretenir une collaboration active avec les médecins*. Its members guarantee: (1) to further research into the cerebral palsy problem, particularly with regard to prophylaxis, improved early diagnosis and therapeutic education; (2) to supervise and encourage the training of specialised personnel; and (3) to advise the Association on its urgent problems and publicise the results of its work.

In all these activities one can see the hand of Dr. Guy Tardieu, whose work at the Hôpital Raymond Poincaré at Garches for long-stay patients is well known.

The secretary of the Association is Dr. Daniel Graveleau, 1 rue Huysmans, Paris 6e. He and his colleagues will welcome collaboration from all those interested in cerebral palsy, particularly ourselves.

Barbara Evans

A French View of Speech Therapy in Cerebral Palsy

IN an excellent article¹, published in French in 1958 and based on a course of lectures at two re-education centres in France, Tardieu, Falinski and Muller gave a clear picture of the cerebral palsied child and the problems involved in providing speech therapy for such a case. In these children, backward development in speech is part of a generalised condition of poor communication with the environment, and therefore training of all the senses should accompany speech stimulation. The authors endorse many of Harold Westlake's principles, and the treatment they suggest is compatible with the Phelps method of physiotherapy.

The importance of the mother's part in the early months is rightly emphasised. In a summary of the evolution and development of speech in the normal child it is pointed out that an infant does not progress from simple vegetative vocalisation to the utterance of meaningful words unless the mother expresses her appreciation and delight, encouraging the baby to repeat the random sounds that are first made purely by chance. This is all the more vital for the cerebral palsied child with a motor dysfunction hampering his efforts; but so often his small vocal achievements are ignored and, without encouragement, his desire to speak is not established. 'A child learns to talk as soon as his parents think he can, and his speech

exists and develops only because they bring it into existence.' (Westlake).

Other conditions that may contribute to undeveloped speech are:

- (a) *The emotional factor*—rejection or over-protection by the parents are examples of conditions unfavourable to the beginnings of speech.
- (b) *Backward mental development* as a part of a general functional immaturity; the functional age of the child should be assessed with the help of Gesell's scale and tests.
- (c) *Developmental aphasia* or specific under-development in the area of word perception, and the ability to express in words.
- (d) *Deafness*, which is a common accompaniment of the cervico-brachial type of athetosis of Rhesus origin.
- (e) *Motor disability* affecting the harmonious working of the respiratory muscles, or the efficient movement of the velum, tongue, lips or jaw. Besides insufficiency of maturation in any or all of these groups, there may be any of the disorders of muscle function commonly found in cases of cerebral palsy.

Plans of treatment are suggested under Westlake's three main headings:

(1) Psychological and Social Readiness for Speech

The aim is to improve the child's total communication with the world around him. In daily life, because of his general physical handicap, there is a risk that he

¹ Tardieu, G., Falinski, E., and Muller, C. 'L' éducation thérapeutique du langage chez les enfants infirmes moteurs cérébraux.' *Neuropsychiat. infant. Hyg. ment. Enfan.* 1958, 6, 3-40.

will have everything done for him without his own wishes being consulted. He should be encouraged to make decisions on even quite small matters, and to show all the initiative he can. He must be spoken to in a language suitable for his age, and all the events that occur around him should be commented on and talked about. He should be told simple stories, with mime and gesture, and mimicking of voices to stimulate interest in speech.

He should also be encouraged to look and listen for things, and to notice the feel, smell and tastes of things around him. Unable to explore his environment with hand and mouth, as normal infants do, he must be given special opportunities to develop his sense perceptions; and while he is appreciating each new sensation, a descriptive word should be repeated to him.

Some cerebral palsied children may appear to be deficient in certain sense perceptions, possibly owing to their lack of experience but perhaps the result of a disorder of figure and background organisation. There may be difficulty in picking out significant details from a mass stimulus, and in this case practice should be given in observing single objects against a plain background, in conditions of minimum distraction. Each object should have some distinct material quality and be as clear as possible; as each is shown an appropriate word must be spoken. This will teach the appreciation of essential qualities that can later be compared and differentiated.

(2) Physiological Readiness for Speech

This requires the education of movement and motor skills based on a knowledge of normal psycho-motor development and the special motor disabilities of cerebral palsy.

Relaxation is very important for the athetoid because his speech varies considerably according to the degree of relaxation he attains. For speech therapy he should

be comfortably placed, either lying down or sitting in a relaxation chair or a straight-backed chair made to fit his measurements. The room temperature should be mild, the lighting subdued, and there should be no visual or auditory distractions. These are the conditions most conducive to relaxation. However, so that the child shall not be too conscious of the act of speaking, it is often helpful to draw his attention outside himself, either by giving him some occupation with his hands or by making him watch the movements of a candle flame as he talks towards it. Gently tapping the child's shoulders and neck, turning his head from side to side, or opening and closing his jaw with gentle manipulation, contributes directly to the relaxation of the muscles of respiration and phonation.

It is helpful with athetoids to have some part of their body stabilised by means of leg braces, sandbags or the therapist's hands, so that no involuntary movements there can trigger off movements in other parts of the body that may affect the production of speech. All these aids are eventually intended to help the children attain voluntary relaxation for themselves, and it is necessary that speech and physiotherapists should combine closely in their work of discovering the most suitable way of helping each child.

Relaxation is beneficial for the spastic too, but he will also need other methods to overcome his dysarthria. These should begin as early as possible to avoid the development of superimposed functional dyslalia.

Breathing. Training in respiration is essential for good speech results, and with the athetoids this is best done through general relaxation. The skeletal muscles particularly should be relaxed, by allowing the head to drop back in hyperextension. Active assisted breathing exercises can be given to encourage a regular rhythm; the therapist's hands are used to guide the

movement only, and no forcible apparatus should ever be employed. It is important to develop the thoracic muscles so that they are strong enough to move in conjunction with the diaphragm and reversed breathing is not established. If there is any unwanted movement in the neck and shoulders, these parts should be stabilised, so that control of expiration can be learnt.

In practice, it is the ability to prolong expiration that is necessary for speech, and the child should be trained to blow through his mouth and a great variety of games can be used to encourage blowing. Velar movement is also exercised in blowing. The tongue should at first be exercised through reflex peristaltic movement; small pieces of food are placed on the tongue for the child to swallow gradually larger morsels are used, and placed further forward on the tongue as ability improves. The child's head, at the same time, is tilted gradually further forward until he eventually learns to swallow lying in a prone position. Tongue placement, as a preliminary to articulation, is first elicited at the involuntary level. Pressure with a spoon or spatula will produce resistant reflex movement in the opposite direction. For voluntary movement pieces of toffee are suggested as directional aids. Chewing-gum stuck behind the upper incisors will encourage upward movement of the tip of the tongue, and the back will be raised reflexly if it is touched with a small piece of bread. The child gradually learns to bring these specific movements under control as he is made aware of them. A rubber block may be inserted between the molars during feeding practice to limit the play of the jaw and ensure that the tongue moves independently, so far as possible.

The same methods can be used to teach control of the lips; taking food from a spoon, chewing, biting, and sucking through a straw are all important preliminary exercises for speech.

(3) Direct Speech Training

Cerebral palsied children vary considerably in their ability to initiate and prolong vocal sound. Vocalisation, however feeble, should be praised and encouraged; if the mother has failed to do this in babyhood it will fall to the lot of the speech therapist later. Rather than giving the child sounds to imitate, one should stimulate him to repeat and develop all chance sounds made in the course of play. Singing with him may also help to encourage his voice.

In the passage from non-verbal to verbal language, no attempt must be made to force the child to speak. He may be asked questions that he will want to answer, but must never feel that his words are impatiently awaited. Care should be taken always to find a special centre of interest for the child as a subject for conversation, and as soon as he acquires a word he should be encouraged to repeat it day after day.

A purely phonetic approach to improved articulation is not always desirable, since these children may not be capable of the discipline involved. Building up from the involuntary movement stage, by watching in the mirror and helping the action with the hands, is the method suggested by Westlake and recommended in this paper.

Later on, with a more mature child, a scholastic approach may be attempted, and he may be given words to learn in a notebook. Later still the improvement of verbal expression and sentence construction may be needed. The authors emphasise, however, that even when the stage has been reached when obedience can be expected, it is still important to alternate periods of learning with periods of play. The chief aim is for the child to grow up into a self-reliant personality, and to this end he should be allowed freedom in his childhood, and be guided through life rather than crushed by excessives cares and demands.

BARBARA CLARKSON

OBITUARY

William J. Bishop

WILLIAM BISHOP carried great learning lightly. In innumerable services he appeared a quiet self-effacing man with an uncanny knack of producing answers from thin air and giving information in some little jest. Indeed all too often, almost before one realised that the jest contained the answer, Bishop had gone, unthanked, yet never leaving one unsatisfied. He had always time for the other man's interests and enthusiasms. He would quote Captain Cuttle's maxim 'When found, make a note of'; and in his 'notes' he always had in mind the diverse interests of a wide circle of friends.

His personal humility did indeed somewhat cloak, at first meeting, his devotion to learning and the strength and vigour which he brought to bear so notably on the advancement of the work of medical libraries and the study of medical history. To gain library experience under his guidance and to work with him was a rare privilege. He set a high standard; his guidance was unobtrusive, his patience endless and his kindness and encouragement unflinching.

He faced, with the same deceptive ease, some minor bibliographical complexity and such a great burden as that thrust on him in the early days of the war. Bishop was then sub-librarian of the Royal Society of Medicine, and at a time when his own home had been destroyed by bombing, and when the then librarian of the Society was incapacitated by a severe and prolonged illness, he met superbly all the administrative problems of maintaining an effective library service without interruption.

Bishop's library career began in the

London Library. From there he went, in 1924, to the Royal College of Physicians; in 1934 he became sub-librarian of the Royal Society of Medicine where he remained until 1946. He then became librarian of the Wellcome Historical Medical Library and in the following years prepared that great collection for its public opening in 1949. He retired from that post in 1953 to give his whole time to medical bibliography and history. Several other libraries have good cause to remember him with gratitude as a consultant and adviser.

It would be impossible to list the books and articles which were enriched by his help, and his editorship of *Medical History* was a potent force in the first British journal devoted to the history of medicine. His own publications, both books and articles, were models. Those, and other works which were approaching completion, will stand as his permanent memorial.

John Shaw Billings, that great librarian, is said to have replied to a compliment on his ability to get work done, '... there's nothing really difficult if you only begin—some people contemplate a task until it looms so big, it seems impossible, but I just begin and it gets done somehow.' Bishop began, and carried through, many things; his death leaves some uncompleted, but his beginning of them is the surest guarantee of their successful completion.

He was a brave, good and learned man, and it is fit that we should remember him with the respect, the admiration and the affection which he himself bore towards the great scholar-physicians who were his own heroes.

P. WADE

Dr. E. Clayton-Jones writes: I wish we could have seen more of Mr. Bishop at the meetings of the Editorial Board. I had known of him vaguely, as one of the almost mythically efficient staff of the R.S.M. Library, since I joined *The Lancet* staff back in 1938, so it was a comfort, in the early days of the *Cerebral Palsy Bulletin*, to know that we had him to advise us on library matters and see that our references were accurate and in proper 'style'. When he came, unofficially, to an Editorial Board meeting for the first time it was a surprise to find that, far from being a dry-as-dust bookish type, interested only in the more abstruse aspects of medical history, he was both jovial-looking and jovial-seeming—able to give a firm, clear and wise opinion on such questions as whether it was easy or advisable to change the name of a medical journal, but also quick to see a heavy joke and cap it with a lighter one. He was an ideal member of a committee such as ours, combining a wide experience of medical libraries and medical journals with much natural wisdom.

We have been glad to find that Mr. Bishop revealed the mysteries of his craft to Miss Sue Goldie, who will be taking over an increasing share of his work for us. When asked for her views on Mr. Bishop as a 'boss', Miss Goldie said he was tremendously kind, considerate and understanding, with a sense of humour that made him great fun to work for; he loved jokes, and the bluer the joke the better. His outside interests were purely literary; he was a faithful fan of Dr. Johnson and a faithful member of both the Johnson and the Osler Clubs. He was immensely widely read, especially in 17th- and 18th-century literature, though his reading was by no means confined to history, let alone medical history. As an illustration of his encyclopaedic knowledge, Miss Goldie recalled his uncanny ability to finish off in five minutes the puzzle on which an ordinary mortal had struggled all the evening.

LETTERS TO THE EDITOR

The Blood-Brain Barrier

SIR—Having read your opening editorial of August, 1961 (vol. 3, pp. 311-314), we feel that the views expressed by Dr. DOBBING on the mechanism of kernicterus should not be allowed to pass without comment. Provocative statements, however heretical, may stimulate fresh thought, but we consider that a number of his statements are refuted by observations which he does not cite. Moreover, the consequences of his views might even be dangerous, because, although he does not directly state this, he implies that the incidence of kernicterus may not be reduced by exchange transfusion.

In his opinion, the old 'primary damage' hypothesis, incriminating anoxia or other noxious agents as the fundamental cause of kernicterus, may have been prematurely dropped in favour of the hypothesis that bilirubin is the cause of brain damage. The reasons for this point of view are difficult to follow, especially as Dr. DOBBING admits the relationship between the incidence of kernicterus and the levels of serum-bilirubin. His hypothesis does not explain why many newborn babies with hyperbilirubinaemia, who have had no evidence of anoxia, have developed kernicterus, or why kernicterus can be prevented by exchange transfusion. He states equivocally that kernicterus 'appears' to be averted by prophylactic replacement transfusion. Does he imply that he is unconvinced by the evidence? The Medical Research Council trial¹¹ showed that in babies with haemolytic disease of the newborn death due to kernicterus was significantly less common in those treated by exchange transfusion (4 out of 62, or 6 per cent) than those treated by simple transfusion (18 out of 57, or 32 per cent). Hsia and others⁸ found that kernicterus was likely to occur in babies with serum-bilirubin levels above 30 mg. per 100 ml. and unlikely to occur when the serum-bilirubin remained below 20 mg per 100 ml.

Dr. DOBBING states that it is no longer believed that there are differences between adult and neonatal brains in their relationships to blood-borne dyes. In support of this he cites the experimental studies of MILLEN and HESS,¹⁰ who were unable to demonstrate macroscopic staining of the brain substance of rats when trypan blue and hydrogen-ion indicators were injected subcutaneously or intraperitoneally. But he fails to quote their comment: '... it may be, however, that the barrier is less complete in the young animal and that its permeability depends to some extent upon the substances used to test it.' For instance, it has been demonstrated that with increasing maturation there is a progressive decrease in the permeability of the blood-brain barrier of the foetus and newborn animal to radioactive phosphorus.¹ NICHOLAS and THOMAS¹² were unable to label the brain cholesterol of adult rats even with massive intraperitoneal doses of ¹⁴C—labelled acetate, but they were able to do so in immature rats. ROZDILSKY¹³ produced kernicterus in healthy newborn kittens by injecting bilirubin-albumin solution intravenously, whereas in puppies and rabbits it was necessary to damage the brain by insulin before kernicterus could be produced. Moreover, KUSTER and KRINGS⁹ produced kernicterus in young rabbits without previous brain damage (anoxic or otherwise), by injecting bilirubin into the carotid arteries. It has been shown in the newborn baby¹⁴ and in rabbits⁸ that the ratio between the concentration of bilirubin in the cerebrospinal fluid and in the serum progressively diminishes with decreasing weight.

Infants with hyperbilirubinaemia, regardless of its cause, may develop kernicterus¹⁹. Of particular interest are the cases of congenital, familial, non-haemolytic jaundice described by CRIGLER and NAJJAR.² These children suffered from a genetic block in bilirubin excretion and had serum-bilirubin levels of 25-45 mg. per 100 ml., without any anaemia or evidence of anoxia. Of seven cases, all but one developed kernicterus and died. Liver-function was normal, apart from delayed excretion of bilirubin. Kernicterus has also been observed in association with hyperbilirubinaemia in congenital spherocytic anaemia,^{5, 7} in neonatal hepatitis, and in cytomegalic inclusion disease.³ Kernicterus also occurs in Gunn's strain of rats which have an inherited deficiency of glucuronyl transferase (the enzyme which conjugates bilirubin with glucuronic acid to form direct bilirubin). In these rats there is a good correlation between kernicterus and hyperbilirubinaemia. Nearly all the deaths with kernicterus occurred in animals with bilirubin levels above 10 mg. per 100 ml.⁴

The yellow pigment in kernicteric brain has been shown to be bilirubin¹². It has been shown that bilirubin is a cytotoxic agent and that it produces a greater fall in oxygen consumption in newborn brains than in adult brains.¹⁴ ZETTERSTROM and ERNSTER,¹⁴ found that bilirubin uncouples the oxidative phosphorylation of brain mitochondria. It is reasonable therefore to postulate that, once bilirubin penetrates into the brain cells, it will exert a cytotoxic effect and will interfere with the emanation of energy from aerobic respiration.

We agree with Dr. DOBBING that the precise mechanism whereby the non-toxic bilirubin-albumin complex gains access to the brain cells is not understood. This difficulty has been stressed by ODELL,¹⁴ who points out that the solubility of free bilirubin in an aqueous medium at pH 7.4 is not more than 0.1 mg. per 100 ml., while a concentration of 20 mg. per 100 ml. is needed for cytotoxicity in isolated mitochondria. ERNSTER⁴ has made the following suggestions to account for this discrepancy: (a) brain function is extremely sensitive to agents acting on cellular respiration; (b) complete inhibition of mitochondrial respiration and phosphorylation is almost certainly not needed to interfere seriously with the activity of the central nervous system; and (c) the brain may possibly concentrate bilirubin by transferring it from the albumin to the cellular proteins. Similarly, the selective staining of various parts of the brain with bilirubin cannot be explained. Nevertheless, we would like to stress that an inability to do so is not evidence against bilirubin being primarily responsible for kernicterus.

In summary, it is established that newborn babies with hyperbilirubinaemia due to a number of different causes develop kernicterus, and that the incidence of kernicterus in infants with haemolytic disease of the newborn can be significantly reduced by exchange transfusions.

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PHILIP F. BENSON
MICHAEL C. JOSEPH

REFERENCES

1. Bakay, L. (1953) 'Studies on blood-brain barrier with radioactive phosphorus: III, Embryonic development of the barrier.' *Arch. Neurol. Psychiat.* **70**, 30.
2. Claireux, A. E. (1961) 'Pathology of human kernicterus.' In *Kernicterus*, Toronto: University of Toronto Press, p. 140.
3. Crigler, J. F., Najjar, V. A. (1952) 'Congenital familial nonhaemolytic jaundice with kernicterus.' *Pediatrics*, **10**, 169.
4. Ernster, L. (1961) 'The mode of action of bilirubin on mitochondria.' In *Kernicterus*, Toronto: University of Toronto Press, p. 189.
5. Hsia, D. Y. Y., Allen, F. H. jun., Gellis, S. S., Diamond, L. K. (1952) 'Erythroblastosis fetalis: VIII, Studies of serum-bilirubin in relation to kernicterus.' *New Engl. J. Med.* **247**, 668.
6. Johnson, L., Sarmiento, F., Blanc, W. A., Day, R. (1959) 'Kernicterus in rats with an inherited deficiency of glucuronyl transferase.' *Amer. J. Dis. Child.* **97**, 591.
7. Joseph, M. C. (1961) Unpublished observation.

8. Kuster, F., Krings, H. (1950) 'Blood destruction and cerebral damage in haemolytic disease of the newborn.' *Lancet*, **1**, 979.
9. Lee, T. C., Hsia, D. Y. Y. (1959) 'Experimental studies on blood-spinal fluid barrier for bilirubin.' *J. Lab. clin. Med.* **54**, 512.
10. Millen, J. W., Hess, A. (1958) 'The blood-brain barrier: an experimental study with vital dyes.' *Brain*, **81**, 248.
11. Mollison, P. L., Walker, W. (1952) 'Controlled trials of the treatment of haemolytic disease of the newborn.' *Lancet*, **1**, 429.
12. Nasralla, M., Gawronska, E., Hsia, D. Y. Y. (1958) 'Studies on the relation between serum and spinal fluid bilirubin during early infancy.' *J. clin. Invest.* **37**, 1403.
13. Nicholas, H. J., Thomas, B. E. (1961) 'Cholesterol metabolism and the blood-brain barrier: An experimental study with ¹⁴C—sodium acetate.' *Brain*, **84**, 320.
14. Odell, G. B. (1959) 'The dissociation of bilirubin from albumin and its clinical implications.' *J. Pediat.* **55**, 268.
15. Rozdilsky, B. (1961) 'Experimental studies on the toxicity of bilirubin.' In *Kernicterus*, Toronto: University of Toronto Press, p. 161.
16. Waters, W. J., Bowen, W. R. (1955) 'Bilirubin encephalopathy: studies related to cellular respiration.' *Amer. J. Dis. Child.* **90**, 603.
17. —, Richert, D. A., Rawson, H. H. (1954) 'Bilirubin encephalopathy.' *Pediatrics*, **13**, 319.
18. Zetterstrom, R., Ernster, L. (1956) 'Bilirubin, an uncoupler of oxidative phosphorylation in isolated mitochondria.' *Nature, Lond.*, **178**, 1335.
19. Zuelzer, W. W., Mudgett, R. T. (1950) 'Kernicterus: Etiologic study based on an analysis of 55 cases.' *Pediatrics*, **6**, 452.

Minimal Cerebral Palsy

SIR—In the June *Bulletin* (pp. 293–295), Dr. Wigglesworth draws attention to 'minimal cerebral palsy' as a valuable diagnostic category. He pleads for early diagnosis and recognition of these conditions to avoid mishandling by parents, teachers and fellow children and believes that simple but early physiotherapy may produce very considerable improvement in the spasticity of such children. But, as he notes, this improvement could also be an effect of maturation. Both Dr. Wigglesworth and Dr. Walton in the August *Bulletin* (pp. 391–392) also refer to associated perceptual defects as having a serious effect on educability, and both believe that here, too, remedial measures are possible.

It may be of interest that in a different field of severe handicap—imbecility—we now have strong experimental evidence that the common perceptual deficiencies can be ameliorated by relatively short periods of individual training. Moreover, such effects are not transitory but persist even when un-reinforced for a period of a year; retention of this type of learning is thus no great problem. In such children there is a deficiency in the spontaneous learning from life experiences which plays such a vital rôle in normal development. Hence those perceptual processes which one takes for granted as developing naturally in normals, and which provide the foundations for all later cognitive development, cannot be so regarded with imbeciles—they appear to need formal teaching. It is no matter for surprise, therefore, that many current training procedures fail through beginning half-way up the ladder rather than at the bottom rung. Conversely, the imbecile's response to proper perceptual training is often remarkable, and some 9-year-old imbecile children have been brought up to the perceptual ability, in complex discrimination tasks, of trained adults of the same mental grade.

It seems just possible that similar mechanisms are at work in the cerebral palsied child. If so, then the earlier such basic therapy is attempted the better. Above all, we need controlled studies of the effects or non-effects of such procedures as physiotherapy and 'sense training' for different types of handicap.—Yours, etc.

The Manor Hospital,
Epsom,
Surrey.

A. D. B. CLARKE

SIR—Dr. Wigglesworth's letter in the June *Bulletin* and Dr. Walton's in your August issue both touched on the concept of Minimal Cerebral Palsy. It was very gratifying to see professional attention focused on a problem which is so rapidly coming to the surface. The awareness that there are a large number of children with minor or even no motor handicap who yet exhibit perceptual and behaviour disorders due to organic brain damage has sparked off great educational changes in the United States within the last few years. Known in America as Brain-injured, Neurologically Handicapped, or Perceptually Handicapped, these children are receiving, in special classes, a structured, disciplined, reduced-stimuli type of training. Significant transformations in behaviour and learning have been achieved. For some, integration into the regular grades has been accomplished, which probably could not have been expected without the special class experience.

As further evidence of the scope of the problem, we have witnessed a rapid growth in numbers and memberships in parent associations throughout America. This parallels the parent movement in cerebral palsy of fifteen years ago, and it seems certain that this current movement will not need so long a time to achieve similar levels.

Our Association has recently published a handbook, '*Helping the Brain-Injured Child*', by Ernest Siegel, M.A., describing the child in lay terms and detailing the educational techniques used to help him. Interestingly enough, while written for the parent, this handbook has sold mainly to professionals and school libraries.

We are heartened by the attention given to our concept of Minimal Cerebral Palsy by two of your readers, and we would welcome contacts with parent groups or any others wishing to exchange information.—Yours, etc.

New York Association for Brain Injured Children,
180 Madison Avenue, New York 16, N.Y., U.S.A.

IRA BRILLIANT,
Corresponding Secretary

Speech Therapy for Developmental Disorders

SIR—I very much appreciate Dr. Ingram's generous reply, in the August *Bulletin* (pp. 392-393), to the questions I raised regarding his conclusions on the value of speech therapy for retarded speech development. Now that he has clarified the position by giving the age range of the children he was referring to, I am in entire agreement with his strictures on this matter.

Although many of my colleagues do give speech therapy to pre-school children and can justify this procedure, my own policy is to use all possible means of parental guidance and to avoid treating regularly children under 5 years, except in special circumstances. If parents are worried about their small child's speech, they are often sent by their G.P. to a paediatrician or the medical officer in a child welfare clinic. These child specialists usually check on hearing and intelligence, and give the parents reassurance and guidance on general handling. The majority of children improve, as Dr. Ingram states. There are some children, however, whose speech remains defective and for whom the paediatrician decides that more specific advice is required. These pre-school children are referred to the speech therapist.

Usually pre-school children are dealt with in the speech therapy department at one interview. A general case-history is taken, and the child's speech is examined in detail to find out in which area the difficulty lies, and its possible cause. Also, an attempt is made to find a suitable method by which the child may be taught so that the mother may be given specific guidance on how to help, if this is considered to be in the child's best interests.

Follow-ups at 3-6 monthly intervals ensure that the child has either improved satisfactorily, or that further advice or investigations will be forthcoming.

Occasionally a pre-school child may be given a few 'treatment' sessions, (a) if the cause of the disorder and possible methods of treatment are still obscure; or (b) to illustrate to the parent how speech can be taught, since few are gifted with the instinct or knowledge of how to proceed and this cannot always be imparted at one interview; or (c) if the child's speech is difficult to interpret, although he has had a normal environmental stimulus, is of average intelligence, with good hearing, and is about to go to school.

I am glad, however, that Dr. Ingram agrees with the value of speech therapy for the schoolchild. When a child has a speech defect which constantly draws attention to itself, and he is of the mental age of 6 years or more, we usually start treatment on a once-weekly basis. If a marked improvement is not made within two terms, the child may be given a rest for a term or further consultation is sought either from a speech therapist colleague, a psychologist, or an appropriate medical specialist.

The problem as I see it is one of management. A speech therapist with long experience can usually, after examining a child, forecast fairly accurately whether a child is likely to improve quickly or slowly, with treatment or without treatment, in the next 6 months. Consequently she can select for treatment those who need a more technical approach at a time when they are likely to respond. If careful selection of cases and timing of treatment are not applied, much time and energy on the part of patient, parents and therapist will be wasted.

I fully agree with Dr. Ingram's comment on the difficulty of determining how much improvement in speech is due to good patient-therapist relationship and how much to the techniques employed, but is this not true in almost every branch of medicine?—Yours, etc.
The Churchill Hospital
Headington, Oxford

C. E. RENFREW, F.C.S.T.,
Chief Speech Therapist.

NOTICE

Prevention of Mental Disorders in Children

FIFTH INTERNATIONAL CONGRESS OF CHILD PSYCHIATRY

Scheveningen, August 24-30, 1962

THE International Association for Child Psychiatry and Allied Professions, which has its headquarters at The Hague, is holding its Fifth Congress at the neighbouring sea-side resort of Scheveningen. The general topic of prevention will be subdivided into the prevention of three groups of noxious factors—somatic, psychical and social. Plenary sessions will be held every morning, the afternoons being devoted to discussions, in groups of about 15, on: (a) Co-operation in the field of prevention; and (b) Dynamics. The speakers at the plenary sessions will be from Italy, Denmark, France, Switzerland, The Netherlands, Germany, Austria, the United States and Great Britain (Dr. Emanuel Miller). The official languages will be English, French and German, and a simultaneous translation service will be provided at the plenary sessions.

Provisional applications should have been made before October 1, 1961, but further particulars can be obtained from The Secretariat of the Fifth International Congress of Child Psychiatry, c/o Holland Organising Centre, 16, Lange Voorhout, The Hague, Netherlands.

BOOK REVIEWS

Papers on Hemiballismus and the Basal Ganglia (1927-1960)

By J. PURDON MARTIN

Reviewed by Thomas E. Twitchell, M.D.

ON the occasion of the centenary of the National Hospital, Queen Square, in 1960, Dr. Purdon Martin has brought together in this booklet reprints of seven of his papers on hemiballismus (hemichorea) and the function of the basal ganglia. These papers reflect both Dr. Martin's continued interest in the problem of hemiballismus over several decades and the evolution of his thoughts on the functions of the basal ganglia in general.

The first paper (*Brain*, 1927, 50, 637) contains a case-report and review of the published work on hemichorea associated with a lesion of the body of Luys. The syndrome of the body of Luys is established as consisting of:

'A violent chorea of the opposite side of the body, with unusual involvement of the shoulder and hip, and therefore with movements of unusual amplitude. The affection of the face . . . is less than one would expect from the amount of choreic movements in the limbs. Speech and swallowing and respiration are all involved in the chorea. The movements cease entirely during sleep. The tendon jerks of the affected side are usually less than those of the other, indicating a moderate degree of hypotonia. In a number of cases . . . the temperature of the choreic side was higher than that of the other half of the body, and in my case there was some unilateral sweating. In almost every case there has been a mental disturbance—at first chiefly an emotional change (excessive anxiety),

and afterwards loss of memory, confusion and disorientation. There are no signs of pyramidal disease, no loss of superficial sensation or a sense of position, no defects in the pointing tests.'

In all cases but one, death resulted from bronchopneumonia, and Dr. Martin suggested that the tendency to bronchopneumonia might be correlated with respiratory disturbance accompanying the chorea.

The second paper (*Lancet*, 1928, ii, 315) is of interest historically as it notes the first description of hemiballismus to appear in the literature in 1870, the first clinical diagnosis of involvement of the corpus Luysii by Jakob, the 'perfect lesion' in Matzdorff's case of hemiballismus with softening entirely confined to the nucleus of Luys, and finally refers to early experimental work on the subthalamic nucleus.

The third paper (with Alcock, N. S., *Brain*, 1934, 57, 504) describes another case of hemiballismus but without involvement of the face. The lesion itself was smaller than that in Dr. Martin's first case, with a relative sparing of the oral pole of the subthalamic nucleus, thus supporting von Santha's argument for a somatotopical localisation in the subthalamic nucleus. In this paper Dr. Martin also suggested that there was no difference between the hemichorea (hemiballismus) associated with a lesion of the nucleus of Luys and other forms of chorea, except in the intensity of the former, which consequently

produced more marked movements in the proximal limb segments.

Although hemiballismus has usually been associated with lesions of the subthalamic nucleus, occasional reports have indicated the appearance of hemiballismus without involvement of this nucleus. Dr. Martin's fourth paper (*Brain*, 1957, **80**, 1) reports three such cases and provides a possible explanation. In the first patient pathological examination revealed no lesion in the subthalamic nucleus. There was, however, extensive degeneration of the fibres passing across the internal capsule from the posterior half of the corpus Luysii to the globus pallidus, the more anterior fibres being intact. In this case the hemiballismus was interpreted in the light of the experimental work of Mettler and his associates, who showed that although a 'choreoid hyperkinesia' in monkeys resulted from focal lesions in the opposite subthalamic nucleus, in a few instances this phenomenon followed a focal lesion not involving that nucleus. They soon demonstrated the importance of the globus pallidus in the production of this phenomenon, for choreoid hyperkinesia could be abolished by destruction of the globus pallidus or its efferent fibres. This led to the hypothesis that choreoid hyperkinesia resulted from a loss of control of the corpus Luysii on the pallidum and this could conceivably result from interruption of the fibres connecting the subthalamic nucleus and pallidum.

Dr. Martin's second patient exhibited hemiballismus which ceased five days after admission to hospital. Examination of the brain revealed a recent lesion of the globus pallidus extending into the internal capsule, and older lesions around the anterior margin of the globus pallidus spreading into the genu of the internal capsule and laterally into the external medullary lamina. The upper part of the ansa lenticularis was destroyed. Dr. Martin

attributed the hemichorea to interruption of the subthalamo-pallidal fibres in the ansa lenticularis and the cessation of chorea to the more recent softenings in the globus pallidus.

The third case is particularly interesting in regard to Dr. Martin's formulation of the patho-pathology. This patient exhibited hemiballismus of the right side, which ceased following the development of a sudden right hemiparesis. Pathological examination revealed some infarction of parts of the left cerebral cortex but no abnormality of the corpus Luysii and other basal ganglia. There was marked cerebral arterial atherosclerosis, and thrombosis of the left internal carotid artery up to and including the syphon. To explain the hemiballismus Dr. Martin suggested that there was a relative greater ischaemia of the corpus Luysii than of the globus pallidus, and provided an explanation for this based on the pathological changes in the vessels supplying these structures and the probable collateral circulation.

The fifth paper (with McCaul, I. R., *Brain*, 1959, **82**, 104) describes a case of hemiballismus abolished by ventrolateral thalamolysis.

The last two papers present Dr. Martin's views on the functions of the basal ganglia. In the first of these (*Lancet*, 1959, **i**, 999-1005) he considers the function of the basal ganglia in relation to two disorders. First there is hemiballismus which can result from a localised lesion of the subthalamic nucleus. A destructive lesion of itself, however, cannot give rise to 'positive' symptoms, and consequently the choreic movement must depend on irregular impulses from some other structure of the nervous system which from experimental studies appears to be the globus pallidus. The second disorder is the muscular rigidity of Parkinsonism. Here Dr. Martin utilises the pathological evidence suggesting in-

volvement of the cells of the substantia nigra in the Parkinsonian syndrome. Again a 'positive' effect appears to follow a lesion of the substantia nigra, yet it has again been shown that a lesion of the globus pallidus, pallidofugal fibres or their end-station in the ventral nuclei of the thalamus can abolish this rigidity. This suggests that the activity of the pallidum is necessary for the maintenance of rigidity, as it was for hemiballismus. Thus, there are two disturbances of function characterised by an excessive discharge of energy, both of which appear to depend on the integrity of the globus pallidus, hemiballismus apparently resulting from irregular impulses, and rigidity from more continuous excessive discharge. Dr. Martin suggests that the subthalamic nucleus in some way controls and modulates the activity of the globus pallidus and that likewise the substantia nigra must have some inhibitory activity on the globus pallidus. In diseases with striatal involvement, such as Huntington's chorea and Wilson's disease, 'positive' symptoms again ensue and it is suggested that the striatum in some way acts also as an inhibitory mechanism on the globus pallidus.

Dr. Martin conceives of the basal ganglia functioning as a group—that is, 'of a dynamic central ganglion, the globus pallidus, surrounded by three controlling ganglia'. A lesion of one of the controlling ganglia will consequently result in 'positive' symptoms, which should not be regarded

as 'release phenomena' but rather as disturbances of the function of the basal ganglia as a whole. 'Muscular rigidity . . . may be indirectly due to degeneration of the substantia nigra; but it is more directly due to the activity of the globus pallidus, in association with the corpus Luysii and the striatum. Similarly, hemiballismus may be due indirectly to destruction of the corpus Luysii, but it is more directly due to the activity of the globus pallidus as influenced by the substantia nigra and the striatum.'

The second paper (*Lancet*, 1960, 1, 999) discusses the function of the basal ganglia in terms of disorder of postural fixation.

After discussing intention tremor as a disorder of postural fixation, Dr. Martin points out how hemiballismus can also be viewed as a disorder in which the mechanism for fixation is acting wildly and suggests that hemiballismus is due to release of a fixation mechanism dependent on the globus pallidus. In like manner rigidity is considered a disorder of fixation with 'over-action of the tonic fixation mechanism'. Continuing his argument he concludes that since these are disorders of fixation, and also disorders of the basal ganglia, then the basal ganglia are concerned with fixation. The paper concludes with a short discussion of 'negative' symptoms, in the form of grave disturbances of posture following pallidal lesion, and some evidence for a bilaterality of pallidal function.

Speech for the Retarded Child

A Teacher's Handbook

Prepared by the Bureau for Children with Retarded Mental Development and Speech Improvement.

New York: City of New York Board of Education, 1961, p. 98, 75 cents.

In preparing this booklet, the Bureau for Children with Retarded Mental Development and the Bureau for Speech Improvement were assisted in their field enquiries

and workshop discussions by a large number of teachers, speech therapists and educational research workers. Although the introduction expressly states that the

basic approach is that of normal speech development, the material provided is all intended for class teaching, and implies a mental age of at least 6 years.

The booklet consists of 4 parts with an introduction and appendix. The introduction is chiefly concerned with the theory of voice-production and includes a 'self-rating' chart to assist the teacher to evaluate her own speech. (It is not explained how the conscientious self-rater can decide how her voice sounds to other people!)

Part 1 is entitled *Classes for Young Children*. If the foundations of spoken language have already been soundly laid in the home, the exercises described would probably be beneficial, but they assume some familiarity with nursery rhymes and fairy-tale characters, as well as with farm animals and common household objects and activities.

Part 2, *Intermediate Classes*, and Part 3, *Classes in Secondary Schools*, deal with the class teaching of speech skills in social situations for children up to about 15

years and contain much fairly advanced material.

Part 4, *Speech Arts for the Retarded*, deals with story-telling, reporting, choral speaking and 'creative dramatics', and is perhaps the best section in the book.

The appendix provides material for simple speech tests and speech games. The booklet concludes with a tabulated summary of the programme already described. Nowhere is there any indication that treatment of any child with defective speech should begin with a thorough paediatric and otological examination.

A booklet which is the result of the combined effort of so many experts cannot fail to be of value to the teachers for whom it was written. The English teacher of E.S.N. children will find it less useful because it is not entirely in tune with her own training and experience. It will be of limited assistance to teachers of mentally handicapped children as we understand the term in Britain.

MARY D. SHERIDAN

Diseases of the Newborn

By ALEXANDER J. SCHAFER

Philadelphia and London: W. B. Saunders, 1960, pp. 878, £7

A comprehensive book for the paediatrician on the subject of neonatal diseases was needed and now Dr. Schaffer has produced one. Apart from a section on cardiology by Dr. Milton Markowitz he is the sole author of the book which comprises 114 chapters in about 850 pages, and some of these in small print.

The British reader must admire the immense amount of work which has been put into the preparation of it and can forgive the few shortcomings. He may, perhaps, regret the birth of yet another new word—'neonatology'—but he is sure to be gratified (and a little surprised)

to find so many references to British work.

It is essentially, as its title declares, a book about *diseases*, and the section on the Normal Newborn is contained in three pages. Even the management of the premature infant is allotted little more than two and an appendix. After this there are sections dealing with the disorders of each system of the body in turn, starting with the Respiratory System, which is covered in great detail. The author's method is to describe a condition under the headings of aetiology, pathology, diagnosis, treatment, and then to give

two or three illustrative case-histories with appropriate chatty comments; for example, 'We have no notion as to the cause of this pleural effusion', or 'Thus far we have been unable to convince our surgical colleagues that this idea has merit, but we propose to persist in the attempt'. One may think this kind of thing long-winded, but it does add to the readability of the book.

Some may find difficulty in following the elaborate classification of the pulmonary syndromes and pneumonias, and perhaps even greater difficulty in applying it, although the last chapter of the section does usefully sum it all up under the heading of 'Differential Diagnosis of Neonatal Dyspnoea'. One is surprised to find intrapulmonary haemorrhage treated as a comparative rarity.

The section on Cardiovascular Disorders by Dr. Mankowitz is almost a textbook in itself, spreading over 100 pages but concentrating admirably upon the newborn period. It provides real help to the paediatrician in attempting accurate differential diagnosis of congenital heart disease, giving examples of E.C.G. and X-ray appearances for each.

There is an equally thorough account of

the other body systems and the impression is gained that no disorder which has ever been recorded is left out. There is, in fact, a regrettable lack of discrimination between the common occurrences and the rarities; for example, congenital hypertrophic stenosis of the duodenum, of which two cases have been recorded in the world's literature, is given a separate paragraph with the same kind of type heading as that for congenital pyloric stenosis.

Surgical aspects are covered from a diagnostic point-of-view, but, wisely, no attempt is made to furnish details of operative procedure. One could have hoped for a little more guidance in the matter of replacement of fluid and electrolyte losses in this age period.

The book is exceedingly well illustrated and not one of the pictures is superfluous. Each has some particular purpose in clarifying the subject under discussion. Full references are at the end of each chapter and a useful appendix deals with dosage of drugs.

This is no book for medical students. Paediatricians will undoubtedly find it of great value for reference purposes if they can sort out the wheat from the chaff.

DAVID G. VULLIAMY

The Challenge of Cerebral Palsy

By PAULETTE A. LEANING, B.A.

Auckland, New Zealand: Whitcombe and Tombs, 1960, pp. 93, 9s.

Miss Leaning has tackled a considerable task—to write a short study of the implications of cerebral palsy for teachers and parents. Herself cerebral palsied, Miss Leaning took her B.A. degree and qualified as a teacher, after which she had 3 years' teaching experience in the Cerebral Palsy Unit at Rotorua, New Zealand. In 1956 the opportunity presented itself for a round-the-world trip to study the latest developments in cerebral palsy overseas,

and it was as a result of this that her little book was written.

This book is written especially for teachers, and aims to help each group of people working with cerebral palsy appreciate the difficulties confronting the others. Her book has little to offer the experienced worker, but it makes a useful introduction for students and qualified teachers thinking of entering this field.

Speaking of staffing, Miss Leaning says

without hesitation that 'no teacher can give adequate instruction, guidance and help to a class of even ten children'. This may well be true, though the teacher will usually not have the ten children in her class all the time—some will be away for physiotherapy, speech training and the like, giving the teacher an opportunity for more concentrated effort with the remainder. Of course, it is not all cerebral palsied children who display the learning manifestations characteristic of brain-injury, such as sensory loss, perception difficulties, perseveration, distractibility, loss of body image, acalculia, cross laterality, etc., and without these defects pupils can function very effectively in a group situation and still receive individual attention in basic subjects. A more generous staffing ratio is necessary for cerebral palsied children in the Nursery and Infant School stage, when intensive individual instruction on the basic skills will greatly ease the future work of teachers.

In some cases, the constant encouragement given to cerebral palsied children, and the development of new opportunities for them lead to over-confidence. The more intellectually successful may then become ambitious beyond their physical and mental limitations. This situation needs careful handling in the latter part of school life.

The problem of keeping records of the behaviour of severely physically handicapped children, whether for research or therapeutic purposes, requires serious attention. The teacher often has no time during either classes or remedial sessions for recording pupils' thoughts, answers, explanations or experiences, and in some schools amanuenses are now being

employed for this purpose. This will leave the teacher more time for actual teaching and help to overcome the frustration of these severely handicapped children.

The sections of the book on the therapies are necessarily short. Miss Leaning particularly stresses the importance of speech therapy and training in view of the social importance of verbal communication. Some will disagree with Miss Leaning on the wisdom of giving occupational therapy to all cases of cerebral palsy, and in fact the use of all the traditional therapies in cerebral palsy needs to be questioned. Perhaps the time is arriving when 'therapy' should be given up in favour of a comprehensive scheme of training and management for development and independent living in its widest aspect.

The very brief section on psychological evaluation pin-points a burning problem in this field to which there is no easy 'question-answer' solution. Everyone working in the field should tackle the question of how best to arrive at a satisfactory diagnosis of a particular child—medical diagnosis has proved totally inadequate for the purpose—and, arising out of the complete diagnosis, how to obtain a workable prognosis. A thorough evaluation of assessment done in the early stages could save all professional workers much valuable time.

In drawing attention to these matters and providing points for discussion Miss Leaning's book will serve a useful purpose. Its sincerity, note of urgency and forthrightness make it well worth reading by all new entrants to this field and an excellent basis for discussion by their seniors.

R. A. PEDDER

ABSTRACTS

In collaboration with *Abstracts of World Medicine*, published by the British Medical Association, and with the kind assistance of the Excerpta Medica Foundation *Courier*, and *Obsterical and Gynecological Survey*

Studies on 5-Hydroxyindole Metabolism in Autistic and Other Mentally Retarded Children

R. J. SCHAIN and D. X. FREEDMAN. *Journal of Pediatrics*, March 1961, **58**, 315-320, 25 refs.

It is thought likely that the biogenic indole alkylamine 5-hydroxytryptamine (serotonin) plays some role in neuronal function in the brain, although the nature of this role has not been clarified. In this study, reported from Yale University School of Medicine, the blood levels of 5-hydroxytryptamine (5-HT) and urinary excretion of 5-hydroxyindoleacetic acid (5-HIAA) were determined in 23 patients aged from 6 to 18 years who had been diagnosed as cases of infantile autism on the grounds of severe preoccupation with self and unrelatedness to other people, a history of onset during the first 2 years of life, and the absence of a history of serious motor retardation. In addition 12 mildly retarded (I.Q. 60 to 80) and 5 severely retarded (I.Q. under 20) non-autistic children were similarly examined, multiple estimations being made in each case. Blood was withdrawn in the afternoon, frozen for up to 24 hours, 5-HT extracted with 20 volumes of acetone, and the extract dried *in vacuo* and kept at -18°C . until bioassay on the heart of the clam or the rat uterus in oestrus could be performed.

The mean normal blood 5-HT levels were 0.02 to 0.15 μg . per ml. Of the 23 autistic patients 7 showed values above normal, rising to 0.540 μg . per ml. In both

mildly and severely retarded non-autistic children the mean values were normal, although the severely retarded gave a higher mean value than did the mildly retarded. Loading with tryptophan in 4 of the autistic children produced no consistent change in 5-HT level, nor was the administration of phenobarbitone, 'dilantin' (phenytoin sodium), or chlorpromazine associated with any change in 5-HT levels. The authors state that the 24-hour urinary excretion of 5-HIAA probably fell in most cases within the generally accepted values of 2 to 9 mg. although the values were distinctly higher in 12 autistic children than in 6 mildly retarded children; creatinine values were much lower in the autistic children, indicating greater dilution of the urine. The symptomatology in the 6 autistic children with the highest blood 5-HT levels did not differ from that in the other autistic patients with normal levels, except that, perhaps significantly, none of them suffered from the seizures which are usually a frequent occurrence in autistic children.

(A scattergram shows great variations in the successive values of 5-HT for each subject, ranging from 0.210 to 0.540 μg . per ml. As it is known that 5-HT decays rapidly in brain tissue the possibility exists that this may also be true for blood. The authors do not state the individual times of storage of blood before extraction.)

G. de M. Rudolf

Cerebral Palsy Twins

E. M. RUSSELL. *Archives of Disease in Childhood*. June, 1961, **187**, 328-336, 27 refs.

The author has studied 44 cerebral palsied twins who attended the Edinburgh clinic of the Scottish Council for the Care of Spastics, and has matched them with a control series of hospital-born twins studied by Drillien in 1958. The 44 cerebral palsied twins were extracted from 488 consecutive cases of cerebral palsy, giving an incidence of twinning of 9 per cent, the highest incidence being among diplegics. The fate of the other twin was studied; less than half (45.5 per cent) of the twins of the cerebral palsied group were alive and healthy, compared with 79.6 per cent of the controls. The majority were stillborn (including those delivered as macerated fetuses), died neonatally or within the first year of life, or were mentally handicapped.

In pairs in which one member was cerebral palsied and the other was still-born or had died in early infancy, the ratio of like-sexed to unlike-sexed pairs was greater than would be expected. There was an excess of first-born cerebral palsied twins and second-born stillborn or dead twins. The average birth-weight of the cerebral palsied infants was less than that of their surviving healthy twins and of the control twins. The stillborn or dead twins had the lowest average birth-weight of all groups and were all premature.

The maturity of the cerebral palsied twins, based on the estimated period of gestation, was considerably less than that of the control twins. The incidence of abnormal pregnancy and parturition was no greater in the cerebral palsied group than in the control group. The ages of the mothers at the time of delivery was no greater than expected.

The cerebral palsied infants showed more abnormal neonatal signs than did

their surviving twins and the control twins. The casualty rate among the 9 pairs of probable uniovular twins was higher than among the 25 pairs of probable binovular twins. Three pairs of uniovular twins in which both members survived are described.

The incidence of mental impairment, visual, auditory and speech defects was considerably higher among the cerebral palsied twins than among the controls.

The authors conclude that, in the majority of cerebral palsied twins, the cerebral defects are unrelated to abnormalities of pregnancy and parturition or to maternal age. The most important factor appears to be low birth-weight, due either to multiple pregnancy alone or to a combination of multiple pregnancy and pre-existing foetal abnormality.

M. C. O. Bax

The Reproductive Histories of Mothers of Patients Suffering from Congenital Diplegia

T. T. S. INGRAM and E. M. RUSSELL. *Archives of Disease in Childhood*, Feb., 1961, **36**, 34-41, 19 refs.

This study, reported from the University of Edinburgh, was aimed at discovering any evidence of impaired reproductive capacity in the mothers of children with congenital cerebral diplegia, of whom two series were studied: (A) 78 mothers of diplegic children living in Edinburgh in 1952 and 1953, and (B) the mothers of 200 such children from all over Scotland seen at the Edinburgh Clinic of the Scottish Council for the Care of Spastics between 1954 and 1959. It is noted that this latter group was not truly representative as the children were mostly selected for suitability for a special school which admits only those of relatively high intelligence. (The correlation between the clinical findings and birth weight in this group was recently discussed by Russell (*Arch. Dis. Childh.* 1960, **35**, 548)).

The peak maternal age at the birth of diplegic child was found to fall between 30 and 35 years, compared to a peak age of 25 to 30 years for mothers of all live births in the general population of Scotland in 1951. However in Series A there was a striking excess of mothers under 20, compared with Series B. The mothers were relatively infertile, with birth rates of 129.5 per 1,000 in Series A and 128.1 in Series B, compared with rates of between 149.4 and 155.3 per 1,000 in mothers in the general population of Scotland in 1951, after excluding childless married women.

The time between marriage and the first pregnancy was similar to that in the general population, but subsequent pregnancies were more widely spaced, and there was a striking lapse of time between the birth of the diplegic child and that of the immediate preceding and subsequent siblings, compared with the spacing between earlier and later pregnancies. When the timing of abortions alone was considered it was found that more than half the abortions occurred immediately after the birth of the diplegic child. The pregnancy resulting in the birth of this child was more often complicated than were the mother's other pregnancies, but one-quarter to one-third of all pregnancies showed no abnormality except premature birth. In only 53 per cent of the multiparous mothers in Series A and 50 per cent of those in Series B had their other pregnancies, confinements, and deliveries been completely normal, and 12 per cent of all pregnancies, excluding abortions, resulted in premature births. In Series A the stillbirth rate and infant mortality were not significantly raised, but in Series B they were twice as high as in the general population, while about half the surviving infants had mental or neurological abnormalities, the majority of these being in the diplegics.

Thus more than half the total conceptions failed to produce a healthy child of average intelligence, but there was no significant difference in the proportions of abnormal pregnancies and confinements resulting in the birth of normal siblings and those producing the abnormal child. The menstrual histories of the mothers showed no deviation from the normal, and the use of contraceptive measures appeared to play no part. *Janet Q. Ballantine*

A Longitudinal Study of the Growth and Development of Prematurely and Maturely Born Children. Part VI. Physical Development in Age Period 2 to 4 Years

C. M. DRILLIEN. *Archives of Disease in Childhood*, Feb., 1961, 36, 1-10, 12 refs.

In continuation of her previous study of the physical development of full-term and premature children born in Edinburgh in 1953-5 (*Arch. Dis. Childh.*, 1958, 33, 423) the author now reports the follow-up results in 528 of these children at their 3rd and 4th birthdays. Two-thirds of them had weighed 5½ lb. (2.5 kg.) or less at birth, the remaining third being full-term babies.

At 4 years the mean weights were closely related to the birth weight. In the premature group the greatest weight increments were recorded for those who were smallest at birth, and the lowest for those who weighed between 4½ and 5½ lb. (2 and 2.5 kg.) at birth. At the age of 4 years the prematurely born are still between 1 and 1½ in. (2.5 and 3.75 cm.) shorter than the mature controls. At 4 years the prematurely born children tended to weigh less and also to be shorter than the controls.

Lower increments of weight and height were more common in families with a poor standard of maternal care. Those who were born at or near term with birth weights of 4½ to 5½ lb. were shorter and lighter at 4 years than children of corres-

ponding birth weight born prematurely. Rates of growth were also related to the stature of the parents. *R. S. Illingworth*

The Incidence of Mental and Physical Handicaps in School-age Children of Very Low Birth Weight

C. M. DRILLIEN. *Pediatrics*, March, 1961, 27, 452-464, 11 refs.

This most recent paper in the author's series of valuable follow-up studies of premature babies in Edinburgh (see *Arch. Dis. Childh.*, 1961, 36, 1) is concerned with 49 children whose birth weight was 3 lb. (1,360 g.) or less, born in 2 Edinburgh hospitals between 1948 and 1960, and now aged 5 years or older. The data provided include information concerning educational attainment, school behaviour, and physical defects.

Approximately one-half of these children were ineducable at a normal school because of physical or mental handicaps or both, one-quarter were dull children requiring special education, and one-quarter were of low average, average, or superior ability. One or more physical defects were present in 26 (53 per cent) and 38 (78 per cent) had behaviour problems. Of 30 with siblings for comparison, 22 were relatively retarded. In 5 of 7 pairs of twins the larger twin scored higher in intelligence tests than the smaller one. There was a significantly higher incidence of handicaps in children from the poorest homes. The incidence of severe handicaps was significantly greater among children born between 1953 and 1954, than in those born in 1948 to 1950. This is in accordance with the view that the proportion of handicapped children is increasing, in this case owing to the increasing number of damaged children surviving. *R. S. Illingworth*

The Eyes and Vision in Infantile Cerebral Palsy

A. A. DOUGLAS. *Transactions of the*

Ophthalmological Society of the United Kingdom, 1960, 80, 311-325. 1 fig., 11 refs.

This is a shortened version of the findings of a survey of the eyes and vision in infants with cerebral palsy carried out in Dundee and the counties of Angus and Perth.

Of the subjects studied, 41.6 per cent were normal ophthalmologically. Among ocular anomalies disorders of motility were predominant—a finding in agreement with that of other published series. The numerous cases of squint included an unusually large proportion of divergent cases. Most of the well-recognised causes of squint were encountered. Choroidal and retinal abnormalities included retrolental fibroplasia, colobomata, and disseminated chorio-retinal atrophy. As regards visual acuity, the incidence of markedly defective vision was higher in the more mentally defective and to some extent in the more spastic patients than in others. Many of the ocular abnormalities appeared to be remarkably well compensated for during the child's mental development. It was clear that with severe mental defect any improvement in the patient's vision would make no material difference as regards educability. *J. D. Abrams*

The Ataxic Form of Poliomyelitis.

M. ARTHUIS, G. LYON and S. THIEFFRY. *Revue neurologique*, Oct., 1960, 103, 329-340, 38 refs.

The authors describe, with brief case-histories, a rare variety of poliomyelitis which has occurred in their experience in only 6 out of some 1,500 cases personally observed. The patients were all children, aged from 2 to 13 years, and the predominating features of the disease were ataxia to a degree that prevented the child walking, clonic movements, and in some cases choreiform movements. On the basis of these and other reported cases they state that in this rare form of the disease

there is rarely any paralysis, and complete recovery after 4 weeks or so is the rule. One feature is tremor of the eyes that may last 2 to 3 weeks. The virus of poliomyelitis, usually Type 1, was isolated from all 6 cases. They conclude that this is clearly a type of polio-encephalitis in which the cerebellum is severely affected. The first recorded case of this type was that reported by Medin in 1898.

G. S. Crockett

The Use of Psychopharmacologic Agents in Retarded Children with Behavior Disorders

G. D. LAVECK and P. BUCKLEY. *Journal of Chronic Diseases*, Feb., 1961, 13, 174-183, 19 refs.

This paper reviews the effects of 5 tranquillizing drugs on children with behaviour disorders at the Rainier School, Buckley, Washington. With the exception of one drug (mephenoxalone) a double-blind technique was used, equal groups of children matched for age, sex, and intellectual ability being compared. (It is not stated whether the groups were matched for severity of disturbed behaviour.) Criteria are not given for the assessment of improvement; this was based on the observations of the nursing attendants.

Of 24 children given fluphenazine, 2 were 'markedly improved' and 8 'improved'. One child was 'markedly improved' on the placebo and 4 'improved'. It was noted that the most severely retarded children derived greatest benefit from fluphenazine. The initial dose of 0.5 mg. daily was increased up to 7.5 mg. daily as indicated by the occurrence of side-effects and the child's clinical response.

Meprobamate, 200 mg., and promazine, 25 mg., were given in combination to 27 children, starting with one capsule 3 times a day and increasing as necessary. Although 11 of these children were improved compared with 4 of the controls, side-effects, particularly drowsiness, were

very prominent and improved behaviour appeared to be related to drowsiness. Spasticity, intelligence, and motor ability were not improved.

Deanol was given to 25 children in a dosage of 75 mg., later increased to 150 mg. daily; 9 of these were improved and 10 of the control group. Deanol was not found to improve attention, reaction time, or distractibility.

Mephenoxalone was given to 27 children, starting at 200 mg. every 12 hours and increasing to 400 mg. 3 times a day. There were no side-effects. There was no improvement in behaviour, but 2 out of 15 children with spasticity were able to straighten previously 'tight' extremities and some muscle relaxation occurred in a further 3.

Chlordiazepoxide was given to 14 patients and a placebo to 14 others for 6 weeks, after which the groups were reversed. The dose given was 10 mg. 3 times a day for those weighing under 50 lb. (22.6 kg.) and 25 mg. 3 times a day for heavier patients. There was no improvement in behaviour, nor was there any reduction in the incidence of convulsions among epileptics; in fact, negative and undesirable behaviour tended to increase.

The authors note that these findings do not support previous enthusiastic reports on the various drugs. They point out the difficulties of assessing changes in disturbed behaviour and stress the need for very careful evaluation of new treatments offered for disturbed and retarded children.

Christopher Wardle

The Puff Test: an Attempt to Assess the Intelligence of Young Children by Use of a Conditioned Reflex

J. RENDLE-SHORT. *Archives of Disease in Childhood*, Feb., 1961, 36, 50-57, 15 refs.

Using a standard puff of air, the apparatus for which is fully described, the author has tested the blink-reaction of 155 children of apparently normal intelligence, aged

from 5 months to 7 years, and 74 mentally defective children with chronological and mental ages ranging from under 6 months to over 6 years. A score was obtained by recording the number of puffs required to establish a conditioned reflex, this being recognised by the child blinking on presentation of the puff-gun, although no puff was actually produced.

Of 12 normal children aged 6 months or less 11 could not be conditioned after exposure to 20 puffs—the normal infantile response. Of 49 aged between 6½ and 12 months 15 gave the infantile response, the others requiring up to 13 puffs; of 32 aged between 12½ and 18 months all were conditioned by 11 or fewer puffs with the exception of one who required 19; and of 47 aged between 18 months and 4 years, 45 were conditioned by 2 to 6 puffs, 2 requiring more; lastly the normal adult response of conditioning after 2 puffs was shown by 13 out of 15 children over the age of 4 years. Scattergrams of the results for the mentally defective children showed little resemblance to those for the normal children in regard to chronological age, but some resemblance in respect of mental age. Over the series as a whole no simple relationship between ease of conditioning and age or intelligence could be established, but there was an almost linear relationship between ease of conditioning and age in normal children

up to about 5 years of age. The author suggests that the puff test is of value in estimating the intelligence of young children up to the age of 4, including those with spasticity or deafness, but not of course those who cannot see. It is quick to perform, usually taking only 2 or 3 minutes and never more than 7, and requires the minimum of co-operation on the part of the testee.

G. de M. Rudolf

Note: Madame Dr Kasantcheva, then President of the Moscow Academy of Paediatrics, when in this country several years ago, stated that an illness, such as rheumatic fever, could prevent the establishment of conditioned reflexes. In standardising and in using the Puff Test, it is essential to specify the state of health of the child.—R. C. Mac Keith.

Observations on the Satellited Human Chromosomes

M. A. FERGUSON-SMITH and S. D. HAND-MAKER. *Lancet*, March 25, 1961, i, 638-640.

Nonmetrical Observations on Cranial Roentgenograms in Mongolism

A. F. ROCHE, F. S. SEWARD, and S. SUNDERLAND. *American Journal of Roentgenology, Radium Therapy and Nuclear Medicine*, April, 1961, 85, 659-662.

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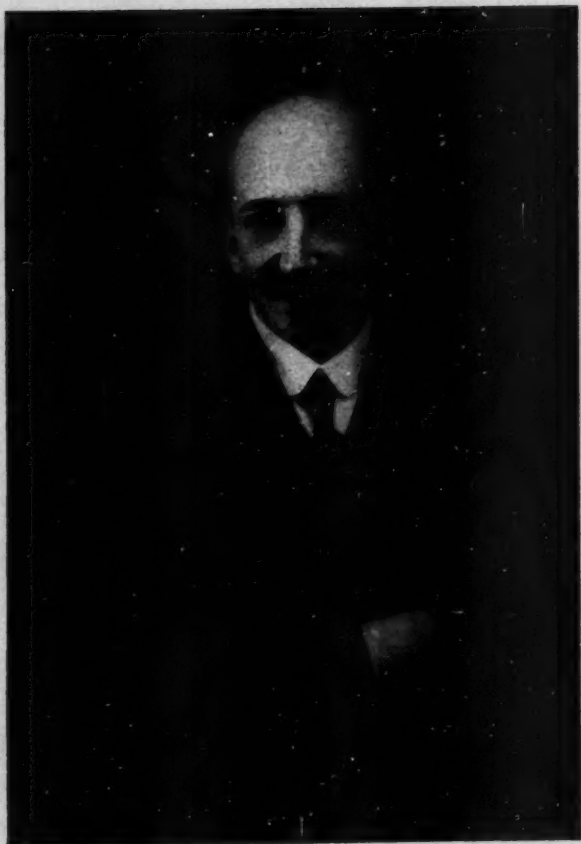
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Our cover picture, by Mr. Graham Clilverd, is reproduced by kind permission of Parke, Davis and Co.



Frederick E. Batten

1865-1918

BIOGRAPHY

F. E. Batten

1865-1918

THE memory of F. E. Batten will always be revered in the history of paediatrics and neurology for in both he was an outstanding pioneer. 'Great Ormond Street' shares with 'Queen Square' the lustre of his fame.

What manner of man was he?

To one who worked under Batten's tutelage close on half a century ago, his personality is still vivid and undimmed. One recalls, as though it were yesterday, the brisk lithe figure with the conspicuous domed head and lively eye, the quick tumbling speech in which the 'r' was only negotiable as 'w', the bubbling humour and the intense interest in current affairs but, first and foremost, in all that concerned the well-being of his patients and those who ministered to them—parents, relatives, nurses, doctors.

Able, enthusiastic, indefatigable—Batten was these in full measure. Yet such dynamic capacity would not have made him so unforgettable had it not sprung from a profound idealism. He sought the truth and ensued it, with unflinching honesty of purpose and with a quiet confidence in his own gifts which begot no trace of arrogance or egotism. His dedication was patently sincere and enriched by a vein of that humility which is the hall-mark of true greatness.

Small wonder that Batten enjoyed the universal respect and affection of his contemporaries. To us of the younger generation he was 'Freddie' and unique.

Inevitably he was a perfectionist, though always practical and purposeful. Notes must be accurate, up-to-date and in typescript, which meant overtime and late nights with the ancient typewriter for the H.P., as I can testify from personal experience. Children loved him and his ward at Great Ormond Street was singularly happy. To his little dark-eyed Scottish Sister, Miss MacDonald, a rigid disciplinarian, 'Dr. Batten' was second only to the Deity. Woe betide anyone who dared to speak to her of 'Freddie'!

The one thing that mattered was to cure *this* patient, or at least to get as near to that ideal as might be humanly possible.

One thrills to recall those frequent consultations—and spirited arguments—with his 'Queen Square' colleagues, Gordon-Holmes and Kinnier-Wilson; those constant spurtings of his surgical confrères to high endeavour, even in the operating theatre where he never failed to attend. Always someone was being goaded to give better than his or her best—pathologist, physiotherapist, pharmacologist, above all patient. The seemingly impossible was only round the corner and the corner *must* be rounded.

These were the pre-duralumin days, when the steel calliper was prohibitively weighty for the flail limb. Hence the research for a material light but rigid, and the birth of the celluloid splint. Under Batten's driving inspiration, the pilot celluloid splints were made by the long-

suffering H.P. The joy of seeing a flail-limbed child up and getting around was ample reward.

There was a risk of inflammability. This was stressed in a topical song at the Christmas entertainment of 1912. Sung to the tune of *The Rajah of Bhong* the relevant chorus ran thus:

Splints, splints, celluloid splints —
They tell me they're very Bon Ton.

If you go near the fire

Then you will not require

Any celluloid splints very long.

Happily there was available the distinguished pharmacist, Mr. J. H. Peck. He was given no peace until he had produced the non-inflammable 'Pexuloid'.

Though neurology was of course his first love, Batten's versatility would seem surprising by modern standards. He took the liveliest interest in the early Rammstedt operations for infantile pyloric stenosis and no one rejoiced more heartily in the successful results. In my time, he had charge of the infectious 'block' at Great Ormond Street, filled largely with cases of diphtheria and pertussis with complica-

tions. His enthusiasm was by no means confined to the occasional instance of post-diphtheritic paralysis.

To Batten, cerebral palsy presented an obvious challenge. The watchword was *re-education* in the broadest sense. The hopeful potentialities were drilled into us all with fervour. Where gross imbalance impeded progress, surgery must be employed to redress it—posterior root section (Förster), obturator blockage, tendon adjustment, etc. Improved mobility was an essential factor in speeding recovery from mental retardation. Such was the Batten gospel—familiar enough now, but in those days a 'new deal' for the 'spastic'.

Came the day in 1918 when Batten, aged 53 and at the height of his powers, had to submit himself to surgery. A prostatectomy was performed and within a day or two he was dead. A tragic and untimely passing. All of us who knew him realised sadly that a singularly bright light had been extinguished, just how bright we can see even more clearly today.

T. TWISTINGTON HIGGINS

Note: With our April, 1962 issue we shall be distributing reprints of two classic papers by Dr. F. E. Batten—'Congenital Cerebellar Ataxia (Cerebellar Diplegia)', from the *Clinical Journal*, 1903, 22, 81-88, and 'Ataxia in Childhood', from *Brain*, 1905, 28, 484-505.—Editor.

EDITORIALS

Human Cytogenetics

THE flood of original papers, reviews and opinions on human chromosomes, many of them conflicting, which have appeared in the medical literature, have presented the clinician with many problems. Are these new techniques of value to him in his day-to-day work or are they merely offshoots of human biological research which from a practical diagnostic point of view are of little value? If this latter viewpoint is correct then he can and will breathe a sigh of relief and continue on his busy way, noting this interesting field of research but from a practical viewpoint ignoring it. It is up to the cytogeneticist and biologist to show the value of their work and explain their results in terms understandable to the clinical mind.

With this end in view a volume entitled *Chromosomes in Medicine** is being published this winter as a *Little Club Clinic in Developmental Medicine*. In this book specialists active in the field of chromosome research describe and review the last ten years' work and attempt to show how these findings can be of value in elucidating some of the problems which confront the clinician. It is to be hoped that this will help by

giving him a new understanding of the problems involved; it may also improve the clinical 'pick-up' of chromosomally abnormal conditions.

In the first chapter SMITHELLS takes the standpoint of the paediatrician and shows what type of case is likely to be of interest for chromosome study. The next three chapters are intended as a background to the clinico-cytogenetic studies reviewed in the last three. First, SYMONDS describes recent work on D.N.A. and ably relates the visible chromosomes to the molecular structure of the genes, describing mutations and chromosome changes from a molecular biological concept. Then HARNDEN describes the processes of cell division, defines many of the terms in regular use in the literature, and deals with the ways in which abnormalities can arise and be transmitted. Finally, the techniques used in this work are described by CLARK. In the next chapter C. E. FORD of Harwell, a pioneer in this field, describes the normal human chromosome set, which we must know about before we can understand the abnormalities.

MURRAY BARR, in a chapter on nuclear sex, describes how the number of X chromosomes can be determined by simple cytological tests and how observations on the interphase nucleus

* Single copies of this book can be ordered from the National Spastics Society, price £2 (or \$6). The subscription to all four *Little Club Clinics in Developmental Medicine* for 1961 is £4 (or \$12).

of buccal mucosa or blood smears can be related to chromosome findings. These tests can be used as a preliminary screening and, in some cases where relationships are now clearly defined, as a definite clinical test to detect some of those cases of sex-chromosome anomaly that interest both the cytogeneticist and the clinician. This chapter will appeal to the biologist, for the biological significance of the sex chromatin is discussed and the question of its origin and relationship to the X chromosomes is aired.

The last three chapters review the major clinical fields to which chromosome studies have contributed. POLANI discusses the sex chromosomes and their abnormalities in relation to clinical conditions in both men and women. Largely owing to the availability of the simple screening method mentioned above, this is without doubt the most extensive aspect of the work at the moment. Sex-chromosome abnormalities are now well known in chromatin-positive Klinefelter's syndrome and chromatin-negative Turner's syndrome and other forms of gonadal dysgenesis. Less well clinically defined are females with three and four X chromosomes.

It is now known that females with primary amenorrhoea frequently have sex-chromosome abnormalities. From a clinical point of view these conditions are defined and classified, and some order is brought into the prevailing chaos. Down's syndrome (mongolism) is described by HAMERTON, and recent advances, particularly in relation to the familial forms of the disease, are discussed. FRACCARO deals with abnormalities not covered by any previous chapters. The other trisomic syndromes are discussed and chromosome studies, in relation to other forms of congenital abnormalities, reviewed.

The purpose of this book is to bring home the importance of this work to the clinician, to explain the jargon and language in use, and so to help him understand the work of the many groups which are now being formed all over the country. If it has achieved at least some of these objects it will have done something towards producing a more dynamic concept of disease and approach to medicine, in which active co-operation between the biologist on the one hand and the clinician on the other is essential.

JOHN HAMERTON

The Effect of Illness and Stress in Pregnancy on the Child

Work by D. H. Stott

IN the first of four papers on this subject,¹ Dr. D. H. STOTT, of the Department of Psychology in the University of Glasgow, explained that his object was to see if there was any link between illness and psychological stress in the mother during pregnancy and mental retardation in the offspring. He studied 103 retarded children, of whom 40 were deemed ineducable and attended an occupation centre and 63 were educationally subnormal with a mean I.Q. of 66. Information about the mothers' health during pregnancy was obtained from 'free-talking interviews', and about the children's early health from interviews, medical files and (in 55 cases) health visitors' records. A control series of 450 children was obtained in various ways. One group comprised sibs of retarded children, another group was made up of 'above-average' children, another of children whose parents were attending adult education centres, and lastly there was a group of the children of professional case-workers. Information was obtained from the parents either by individual or group interviews or from a questionnaire. About a quarter of the adverse pregnancy factors reported were illnesses; the remaining three-quarters consisted of different types of psychological stress. Illness and stress were much more frequent in the retarded than in the control group. In addition to the association between

illness and stress in pregnancy and mental retardation in the children STOTT found an unexpected and significant association between illness and stress in pregnancy and both early ill-health in the children and congenital malformations. He considers these relationships likely to be causal. The possibility that the mothers of the retarded children remembered better or imagined incidents because their children were retarded he discounts for several reasons: (1) that 'old wives' tales' have been killed by family doctors and that contemporary folklore favours hereditary explanations; (2) that the pregnancy factors which had sequelae were such as would have been remembered by parents of normal children; and (3) that the association of the pregnancy factors also with early ill-health and malformation in the child supports their reality.

In a second paper² STOTT analyses the psychosomatic stresses during pregnancy in his original sample, to which are added the findings from questionnaires completed for a very large series of mental defectives. In all, the pregnancy histories of 365 mothers of children with mongolism, and 484 mothers of other mentally defective children are compared with the original controls. Shock, distress, harassment, etc., were all found to be more common in pregnancies antecedent to the birth of

children with either mongolism or other types of mental defect. The timing of the stresses suggests an excess at the end of the first trimester in mongolism, and a deficit of stresses reported in the first trimester both in mental defect other than mongolism and in the controls.

In a third paper³ a correlation, not of a very high order, was observed between adverse pregnancy factors and 'unforthcomingness' in a series of 40 ineducable children, 62 educationally subnormal children and 86 retarded children in ordinary schools. 'Unforthcomingness' was one of six personality types into which the children were grouped. It was the commonest abnormal personality-type in a small random sample of children aged 7 to 11 years in Bristol primary schools.

STOTT wished to test whether the early ill-health was responsible for the retardation or whether it and the retardation had a common origin, in, for example, poor social conditions⁴. An attempt was made to trace 198 children who had been admitted to hospitals in Bristol during 1946 and 1947 for two weeks or more when they were under the age of 2 years; 72 per cent of the children were found at schools in Bristol and each was matched with a control child of the same sex and age attending the same school. The educational achievements of all the children were measured and those who had been admitted to hospital were found to be substantially retarded compared with the control

group. Within the hospital group, scholastic achievement was found to be strongly related to social conditions, but these were not held responsible for the children's backwardness. It seems possible, however, that the home conditions of the children who were admitted to hospital in the first two years of life were inferior to those of the control group and that their educational retardation might be explained by these and associated factors.

STOTT has made a serious effort to examine the possible effects on the child of maternal illness and mental stress during pregnancy. The evidence he has produced so far is consistent with an adverse effect manifesting itself in mental retardation, early ill-health and malformations, but before accepting his opinion that the relationships found are likely to be causal, other possible explanations must be carefully considered. Many would not share his faith that mothers of retarded and malformed children would give an unbiased history of their pregnancy and would satisfactorily remember and time events which happened so long ago. Unfortunately it seems almost impossible to obtain satisfactory evidence on the relationship between pregnancy factors and congenital defects in a retrospective manner, while the cost of prospective surveys, in time, effort and money, may be out of proportion to their results.

ALISON D. McDONALD

REFERENCES

1. Stott, D. H. (1957) 'Physical and mental handicaps following a disturbed pregnancy.' *Lancet*, i, 1006-1012.
2. — (1958) 'Some psychosomatic aspects of causality in reproduction.' *J. Psychosom. Res.*, 3, 42-55.
3. — (1959) 'Evidence for pre-natal impairment of temperament in mentally retarded children.' *Vita hum.*, 2, 125-148.
4. — (1959) 'Infantile illness and subsequent mental and emotional development.' *J. genet. Psychol.*, 94, 233-251.

Hemiplegia in Children and Adults

FROM September 18 to 23 a study group met at Wills Hall, Bristol, under the auspices of the National Spastics Society, to discuss hemiplegic cerebral palsy in children and adults. Once again, as after the two N.S.S. International Study Groups in Oxford, the participants departed at the end of the week with expressions of gratitude for the value of these meetings.

The 77 participants came from America, France, Scandinavia, Switzerland and Italy, as well as this country. The conference worked through all the aspects of hemiplegic cerebral palsy. The first paper was on aetiology by Dr. CORNER, who gave an account of her survey and follow-up study of premature babies with neonatal disorders, and the last paper was on the employment problems of hemiplegic patients with cerebral palsy by Miss M. R. MORGAN, who has a wealth of experience in this problem in her capacity as Employment Officer of the National Spastics Society.

Perhaps the three outstanding features of the week were as follows. The first was the clash of views on the value of physiotherapy. The vigorous discussion on this subject was initiated by Dr. HUME KENDALL's paper in which he reported the formation of a research project to investigate the effects of physiotherapeutic treatment. Welcoming this activity, Dr. J. P. M. TIZARD clearly stated his own rather heretical views on the problem; his feelings

were that, while any form of treatment in the broadest aspects might help the cerebral palsied patient, he himself was doubtful whether physiotherapeutic measures were actually of value in hemiplegics. Inevitably, this bold statement met with a great deal of opposition from the group in general, who felt that many of their number had much clinical experience to offer on the value of such treatment (Mrs. BERTA BOBATH, for example, gave a clear account of her aims in the treatment of this condition). The resulting discussion did, however, emphasise the need for controlled clinical trials of all forms of treatment—a point which was reiterated by the surgeons present when the conference discussed the surgical treatment of hemiplegic cerebral palsy.

A great deal of attention was paid to the accompanying psychological disturbances. One had the happy feeling that at last the psychological aspects of cerebral palsy were beginning to get the attention they deserved. A particularly interesting paper in this group was presented by Dr. CHRISTOPHER OUNSTED, who gave an account of intellectual disabilities in relation to lateralised features in E.E.G. recordings. In his long, complex, careful paper he described a detailed study of 109 children with normal and abnormal E.E.G.'s and related these studies to scores in verbal and non-verbal psychological tests. The results gave rise to a number of controversial thoughts about the

need to revise our concept of a 'dominant' hemisphere.

Emotional as well as psychological problems received attention; Miss E. F. TOWNSEND drew attention to various periods of crisis in the family of the handicapped child and Dr. R. C. MAC KEITH discussed *The Doctor, the Child and the Family*, laying especial emphasis on the prophylactic value of deep understanding and close support in the period immediately following the recognition of the existence of a handicap.

One of the advantages of meetings such as this is the chance they give people from other countries to discuss their common problems informally. Dr. A. MILANI-COMPARETTI was anxious to discuss with his British colleagues some revision of the Little Club classification of cerebral palsies, which he has been using in Italy and finds a most valuable aid whose common use he would like to see extended. On two evenings a dozen people met to discuss this classification and they considered some minor changes which are to be circulated for comment.

The value of such study groups is very clear to the participants at the time, but if they are to have any lasting value, and particularly if they are to help those who did not attend the discussions, the ideas and thoughts presented must be published for a wider readership as quickly as possible. The usual lengthy delay in the publication of the proceedings of such conferences seriously reduces their usefulness. The organisers of the Bristol conference were determined that their proceedings should be printed before the impetus which the participants had derived from the meeting had died down. The papers have, therefore, already been published, two and a half months after the meeting, as a *Little Club Clinic in Developmental Medicine*, which has been circulated to subscribers and is obtainable through the National Spastics Society.* The editors are somewhat proud of this feat of medical publication.

M. C. O. BAX

* Hemiplegic Cerebral Palsy in Children and Adults. *Little Club Clin. dev. Med.*, 1961, No. 4. Price £1 (\$3) from N.S.S., 12 Park Crescent, London, W.1.

ORIGINAL ARTICLES

On Postural and Righting Reflexes

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'Posture follows movement like a shadow; every movement begins in posture and ends in posture.'—C. S. SHERRINGTON.¹⁴

IN order to understand the disordered muscular activity of cerebral palsy, a knowledge of the postural reflexes is essential. The postural reflexes are automatic reactions which maintain the characteristic orientation of the body with respect to gravity, both during movement and at rest. Certain sensory organs are responsible for signalling any deviation from the normal posture, and the most important ones are appropriately situated within muscles themselves¹³, within the muscles and joints of the neck, and within the head (the labyrinth of the inner ear). When a normal voluntary movement is begun, the postural reactions accurately adjust themselves; the movement arises out of a posture which is itself gradually and sufficiently inhibited in the prime movers and antagonists to allow smooth and co-ordinated movement to occur, until finally, on ceasing the movement, a suitable posture is re-attained. Meanwhile, more general postural reflexes have maintained body balance as a whole and an attitude most suitable for visual control of movement, if need be.

One of the disorders of function in cerebral palsy is a failure of the correct interaction of postural reflexes and voluntary movement, a failure that is partly due to a loss of the higher inhibiting mechanisms which normally damp down

the postural reactions and allow movement to be superimposed on them. This loss of cerebral inhibition may cause the postural reflexes to be greatly exaggerated in cerebral palsy, and may reduce the patient's movement almost to a marionette state, the guiding force being gravitation. Voluntary activity may then be very restricted, but the patient can be taught how to initiate postural adjustments of limbs which are not themselves under direct voluntary control. This is an interaction of local and general static reactions which will be discussed later.

Much of our knowledge of postural reflexes stems from the inspiration of Sherrington and his pupils, Denny-Brown and Liddell in Oxford. It was they who contributed so much on the basic reflexes of the spinal cord and their linkage with posture. Since then, physiologists of many nations, including Magnus, Rademaker, de Kleijn, Schoen, Barraquer-Bordas, André-Thomas and Bard, have investigated successively higher centres of the nervous system, in which more general and often dominating postural reactions are organised and integrated with the spinal reflexes. Animals such as cats, dogs, rabbits and monkeys were used to elucidate these various reflexes, but there is no doubt that in man the postural reflexes follow a similar pattern and distribution of organisa-

tion, and are of a similar nature. In quadrupeds the head is the leading segment, bearing special sense organs which can register events at a distance—namely, the nose, ears and eyes—and it is hardly surprising that the postural reactions of the limbs and body are subjugated to the position of the head in relation to them.¹² Though in man the head is not usually the leading segment, the presence there of the eyes—now the dominant distance-receptors—endow it with even more special significance, particularly in the performance of skilled movements under binocular visual control.

For the sake of convenience the postural reflexes will here be discussed under subdivisions determined by their centres of organisation, but it must be remembered that there is interaction between these centres in the various levels of the nervous system to maintain balance and posture as a whole and to allow smooth and co-ordinated voluntary movement against this background.

THE SPINAL LEVEL

The isolated spinal cord has a large repertoire of limb reflexes including postural and stepping reflexes, yet a spinal animal is unable to stand unsupported and cannot initiate nor maintain walking. It lacks the organised facilitatory and inhibitory balance provided by higher levels of the nervous system.

1. Local Static Reactions of the Spinal Cord

These are confined to a single limb and are concerned with standing, though in a wider setting they provide the background for voluntary movement and the co-ordination of muscles.

The Stretch Reflex.—When a muscle is stretched, very sensitive sensory organs within the muscle (muscle-spindles) are stimulated, and they discharge a volley of impulses to the spinal cord. Here they excite, with the least possible delay,

motoneurons of only that muscle which was stretched. A stretched muscle, then, tries to contract in order to prevent lengthening, and this is the stretch reflex.¹³ Some muscles normally show a well-marked stretch reflex, and these are muscles which, in the normal attitude of the animal, oppose the force of gravity.

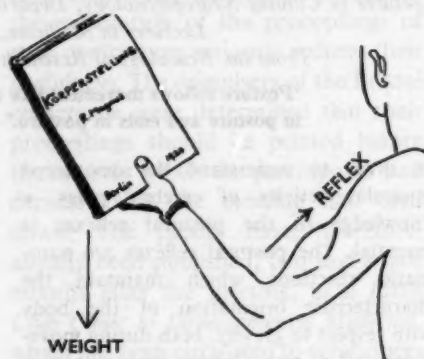


Fig. 1. A stretch reflex.

The weight of a limb will stretch certain muscles, and their lengthening will tend to be prevented by the stretch reflex of these muscles (Fig. 1). Similarly, the weight of the body and head will stretch muscles connecting the limbs with the body and the head with the neck, as well as the extensor muscles of the back, and stretch reflexes will be evoked in all these, thus fixing posture against gravity by means of a single local reflex in each individual anti-gravity muscle.¹⁴

The stretch reflex is well designed to maintain posture for long periods, for muscle-spindles do not readily fatigue but will discharge whenever the muscle is lengthened. Furthermore, anti-gravity muscles contain a large proportion of small, dark-red muscle fibres which are packed with myoglobin (an oxygen-carrying pigment), have contraction times of about 1/10 second, and may be fully tetanised at less than 30 impulses per

second. This is a great economy in comparison with the paler, faster muscle fibres found predominantly in the muscles concerned with eye movements, running and other swift alternating movements, for they may require 100-300 impulses per second to tetanise them and they can maintain their tension only for a very short time.⁶

It should be mentioned that the discharge of muscle spindles has a wider local effect than that described above, for not only does it excite its own motoneurons but it also inhibits the motoneurons of the antagonistic muscles. This is the basis of reciprocal innervation—a necessary accompaniment of all alternating movements.

The stretch reflex is a simple monosynaptic spinal reflex which is, nevertheless, limited and controlled from other centres, within the brain. This is through descending pathways which either inhibit or facilitate the motoneurons themselves, or, less directly, the gamma motoneurons. The latter innervate small muscle fibres within muscle spindles which allow an adjustment of the sensitivity of this sensory organ and thus have control over the intensity of the discharge in response to stretch.^{7, 12}

The Positive Supporting Reaction.—This consists of simultaneous contraction of opposing muscles so as to fix joints.^{10, 11, 13} It is not designed for the prolonged maintenance of posture, but is the basis of such attitudes as the poise before springing or standing to attention. It is a reflex capable of supporting a great weight, and is particularly marked in the hind limbs. The stimulus for the reflex is a combined one of tactile and proprioceptive components (stretching the toe muscles), resulting from pressure on the sole of the foot near the toes.

This reaction is completely inhibited by plantar flexion, so that the limb can be

rapidly transformed from a supporting pillar to an active moving member.

2. Segmental Static Reactions of the Spinal Cord

These reactions provide a wider co-ordination of posture between all four limbs and the body.

The Flexion and Crossed Extension Reflex.—This provides support and the maintenance of balance whenever a limb is withdrawn from an injurious agent (Fig. 2). The noxious stimulus is more effective if it is applied to the sole or to the palm, but reflex flexion of the limb, as a result of stimulation of the limb at other sites, may occur if the stimulus is strong enough. The extension of the contralateral limb, in the intact animal, may be strengthened further

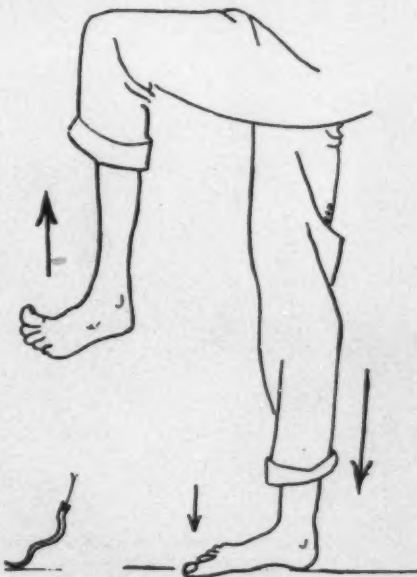


Fig. 2. The reflex of flexion and crossed extension.

by the positive supporting reaction. The reflex of crossed extension long outlasts the brief stimulus which excites the flexion reflex.

The reflex pattern of flexion and contralateral extension may provide one of the phases of reflex stepping, though here the stimulus is clearly of a different nature.

Long Spinal Reflexes.—These co-ordinate the relative postures of the fore and hind limbs, and they assume great importance during quadrupedal walking.

3. General Static Reactions of the Spinal Cord

These reflex responses are more widespread than the ones already mentioned, and they determine the posture of all four limbs and the body in relation to the relative position of the neck.

The Tonic Neck Reflexes.—These are integrated in the upper cervical segments of the spinal cord and are very powerful

Magnus and de Kleijn, who described five components. When the neck was rotated, the fore limb, on the side pointed to by the jaw, tended to extend, while the opposite fore limb flexed (Fig. 3). Bending the neck towards one shoulder produced the same result. These are the so-called asym-



Fig. 4. The reflex effects on the limbs of extension (dorsiflexion) of the neck.

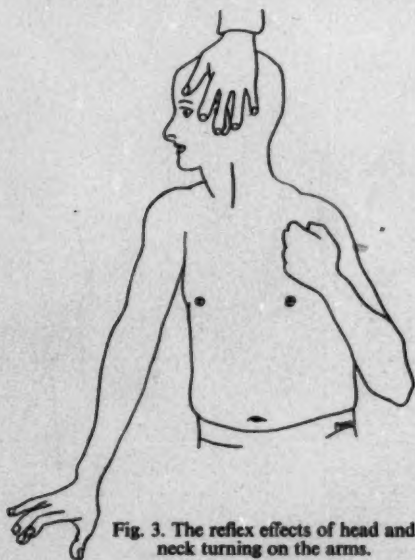


Fig. 3. The reflex effects of head and neck turning on the arms.

mechanisms for co-ordinating the limb postures with the neck.^{10, 11} The reflexes depend on sensory impulses which are derived from the muscles and joints of the neck. They were intensively studied by

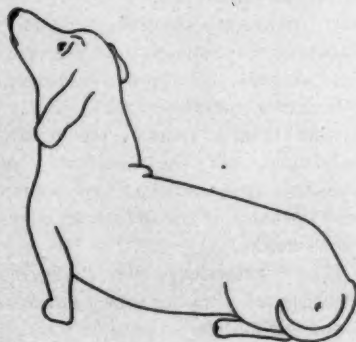


Fig. 5. The reflex effects on the limbs of flexion of the neck

metrical tonic neck reflexes. When the neck was dorsiflexed, both fore limbs extended and the hind limbs flexed—the attitude of an animal looking upwards (Fig. 4). Flexion of the neck produced flexion of the fore limb and extension of the hind—the posture assumed by many quadrupeds while sniffing the ground (Fig. 5). These, like the next, are symmetrical tonic neck reflexes. Finally, some quadrupeds show a 'vertebra prominens' reflex which consists of the flexion of all four limbs in response to a pressure stimulus in the region of the lower cervical spine. This clearly serves to adjust the posture of a quadruped while passing under an obstruction.

Sensory impulses from the neck can also produce compensatory movements of the eyes in such a way as to secure central binocular vision in the direction of turning.

THE POSTURAL REFLEXES OF THE MEDULLA

The decerebrate animal, in which the brain above the medulla has been destroyed, shows all the postural reflexes that we have so far described, though in an exaggerated state due to the loss of inhibitory control from the brain.¹⁵ It can stand unsupported, although it cannot right itself if displaced, and, in addition, it exhibits further general static reactions which are derived from sensory impulses originating in the otolith organs in the labyrinth of the inner ear. An animal in its natural posture, with its head at 45° snout down, has minimal labyrinthine effects on its four-limb musculature; but if it is placed in the supine position, with the head at 45° snout up, then maximal extension of all four limbs occurs in response to the otolith stimulation.^{16, 17} Intermediate positions of the head, between the two extremes described, are thought to have intermediate effects on the extensor musculature.

In addition to these effects on limb musculature, the neck muscles are excited

or relaxed by impulses arising in the otolith organs. Neck turning so produced may then secondarily enforce postural changes in the limbs through the tonic neck reflexes.

Some influence on the eye musculature is also exerted from the otolith organs to ensure that the visual fields are kept reasonably constant.

THE CEREBELLUM

It has been generally believed that the cerebellum is little concerned with the postural reflexes, and this may well be so in acute experimental animals undergoing cerebellectomy. However, the profound effect on postural reflexes of chronic cerebellar lesions in man and animals, can leave no doubt that the cerebellum plays an important rôle in a general way.^{8, 18} In more recent years, it has become clearer that the cerebellum may provide a centre for the co-ordination and interaction of postural reflexes and voluntary movements in relation to sensory impressions coming from moving muscles, the eyes, and possibly other sense organs.⁷

THE MIDBRAIN AND THALAMUS

An animal whose brain has been destroyed as far back as the thalamus can stand in a normal posture and shows appropriate compensatory postural changes on turning the head and neck. In addition, it can now right itself if pushed over.

Righting Reflexes

The sites of the centres within the nervous system, concerned with the elaboration of the righting reflexes, have not been adequately worked out. However, the sensory impulses which determine the outcome are known to originate in the otoliths of the labyrinth, from tactile receptors in the trunk in particular, from the neck and from the eyes. The optical righting reflexes will be considered later as

they depend on the occipital cortex of the cerebral hemispheres.

Labyrinthine Righting Reflexes.—Provided the otolith organs of both labyrinths are normal, then the head will be righted with respect to gravity, irrespective of the position of the body and independent of other sensory impressions.^{10, 11} Since movements of the head are linked with neck movements, which by the powerful tonic neck reflexes influence limb and body posture, the labyrinthine head-righting reflex is of great importance in restoring head—neck—limb—body posture with respect to gravity.

Body Righting Reflexes Acting on the Head.—Asymmetrical stimulation of tactile or pressure receptors on one side of the body (such as the weight of the body lying on a table) can provide the cue for the muscular sequence which results in righting of the head in a labyrinthectomised animal. Under normal conditions this reflex must facilitate the labyrinthine righting reflexes.

Neck-righting Reflexes.—Impulses arising asymmetrically from the neck muscles in a rotated position, result in compensatory reflexes which tend to right the body in relation to the head.

POSTURAL REFLEXES DEPENDING ON THE CORTEX

It is currently believed that in normal primates and in man the postural reactions of lower levels of the nervous system are controlled and inhibited by the motor regions of the cerebral cortex and their outflow through the basal ganglia.

Optical Righting Reflexes.—These require the mediation of the occipital cortex and probably need reasonable vision and visual orientation in space in the absence of other sensory cues. In normal individuals it probably has a dominant rôle, though facilitated and added to from other receptors.

Placing Reactions.—These consist of

postural adjustments of the limbs in response to a moving tactile stimulus which allows the foot to be placed and weight supported on a solid object. They are usually tested by drawing a blindfolded animal's foot gently over the edge of a table, when the foot will at once be placed firmly and squarely on the table. This reaction depends on both the motor and sensory areas of the cerebral cortex.

Hopping Reactions.¹—These are compensatory movements which tend to bring a limb back from a displaced position. They are largely of tactile and proprioceptive origin, but depend on the motor area of the contralateral cerebral cortex.

STATO-KINETIC REFLEXES

So far we have considered the various mechanisms that maintain posture in the static state or at repose, and the means whereby posture is regained and retained if the animal or subject is displaced. Statokinetic reflexes, however, are produced by movement of the head or limbs and not by an abnormal orientation.

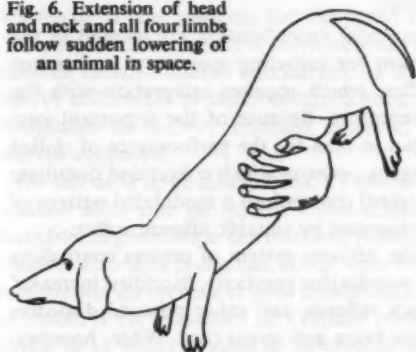
The Reactions of Movements of Limbs

We have already described the reflex adjustments of posture that are possible when a limb is moved either voluntarily or reflexly. There is an interplay of inhibition and excitation at the spinal level (reciprocal innervation), and, depending on the nature of the movement, flexion and crossed extension reflexes, positive supporting reactions, and stretch reflexes may all play an important part in adapting posture to movement.

Reactions to Movement of Progression (Linear Acceleration)

When a quadruped is suddenly lowered through the air, its limbs rigidly extend and its head also extends, even when blindfolded (Fig. 6). This response is similar to

Fig. 6. Extension of head and neck and all four limbs follow sudden lowering of an animal in space.



that which might be seen when an animal lands after jumping from a high place. In man, it is a common experience to notice strong extension of the legs while travelling up in a lift, and slight flexion at the hips and knees while travelling down. Forward progression in man tends to extend arms, legs, head and neck, and backward progression produces the reverse.

Reactions to Rotation (Angular Acceleration)

Reactions of the Head and Eyes.—If the head is rotated about a vertical axis, the head and neck and eyes will turn to the opposite side as if to keep the gaze on a fixed point, but as the body turns further the head and eyes make a quick adjustment as if seeking a new fixation point, and then drift again in the direction opposite to the accelerating movement. This is head and ocular nystagmus, and when the movement ceases there is nystagmus in the opposite direction owing to the time lag in the movement of the labyrinthine fluid on deceleration.

Reactions of the Body.—With rotation around the vertical axis there may be bending of the trunk towards the direction of movement with extension of the limbs on that side and a tendency to flexion on the opposite side.

RÔLE OF PROPRIOCEPTORS WITHIN THE EXTRAOCULAR MUSCLES

From what has been said, the head is all-important as the bearer of the distance receptors, particularly the eyes, and through its proprioceptors (the labyrinths of the inner ear) it exerts a powerful influence over the body musculature. Posture of the head is closely linked with the eye muscles, and compensatory movements are all designed to provide central binocular vision, in a direction of orientation to gravity, which is the usual plane for our visual experiences and interpretation. It is of some interest to enquire whether the eye muscles themselves contribute, by means of proprioceptors, to this postural control. Receptors which respond to stretch have been demonstrated in the external ocular muscles, and fairly simple muscle spindles are to be found in histological sections.¹⁷ There is no local stretch reflex; but instead, the afferent impulses from the stretch receptors are distributed to the motoneurons of antagonistic eye-muscles (possibly not directly but through the superior colliculus). They also pass up the brain-stem to the superior colliculus and even to the cerebral cortex. Reciprocal innervation and strict co-ordinate control of binocular adverse movements may be partly mediated through this sensory influx from muscle spindles, for they are so particularly sensitive (and can be made more or less so by the discharge of gamma motoneurons) that tiny deviations from the visual axis could be accurately signalled. Physiological experiments have, so far, elucidated only a relatively few bare facts on this subject. Some workers, such as von Tschermak,¹⁸ believe that proprioceptors within the eye-muscles are responsible for judgment of visual space, but this view seems to dismiss some of the purely visual aspects of binocular vision and the rôle of previous experience and learning in almost every judgment we make.

CONCLUSIONS

Many postural mechanisms and centres for muscular co-ordination are located in the spinal cord. What is missing there is a mechanism for initiating movement and motor behaviour appropriate to the whole sensory influx, which requires integration with the needs of the head—the bearer of the distance receptors. Because of the important part played by the eye, not only for species survival but in man for the performance of skilled visual tasks, the head and neck contain sensory organs—centres which collect and distribute information and which dominate and mould the spinal centres into a modulated pattern of activity, initiated, controlled, co-ordinated and integrated by suitable afferent influx.

Owing to the widespread distribution within the nervous system of centres controlling posture and tracts interconnecting them, it is little wonder that spasticity, flaccidity, increased positive supporting reactions, increased tonic neck reflexes, and other postural disorders are so commonly seen with almost any lesion of the brain and spinal cord. When, however, the postural disorder is established, it should be studied to enable the level of the lesion to be roughly localised and also to determine what reflex manoeuvres can convert it into predictable, controlled, and therefore useful movement.

The Bobaths have already explored this fundamental approach to the understanding and therapy of cerebral palsy.^{1, 2}

Über Haltungs- und Korrektionsreflexe

Viele Haltungsmechanismen und Zentren für Muskelkoordination liegen im Rückenmark. Was dort fehlt ist ein Mechanismus für den Beginn der Bewegung und des an den ganzen sensorischen Strom angefassten motorischen Verhaltens, welches mit den Bedürfnissen des Kopfes—Träger der Entfernungsrezeptoren—integriert werden muss. Wegen der wichtigen Rolle die das Auge spielt, nicht nur für das Fortleben der Gattung sondern auch, beim Menschen, für die Ausführung spezialisierter Schaufgaben, enthalten Kopf und Nacken die Sinnesorgane, Zentren die Auskunft einsammeln und verbreiten, die die Zentren des Rückenmarks beherrschen und in eine modulierte, eingeleitete, kontrollierte, koordinierte und durch geeigneten afferenten Strom integrierte Tätigkeit anordnen.

Wegen der ausgebreiteten Verteilung, innerhalb des Nervensystems, der Zentren, die Haltung kontrollieren und der Trakte, die sie miteinander verbinden, ist es nicht erstaunlich, das man Spastizität, Schlaffheit, gesteigerte positive Stützreaktionen, gesteigerte tonische Nackenreflexe und andere Haltungsstörungen so oft bei fast allen Läsionen des Gehirns und Rückenmarks antrifft.

Wenn jedoch die Haltungsstörung zustande gekommen ist, muss man sie studieren, um eine annähernde Lokalisierung der Höhe der Läsion möglich zu machen und auch um festzusetzen, welche Reflexmanöver sie in eine voraussehbare, kontrollierte und, folglich, nützliche Bewegung verwandeln können.

Im Bereiche der Anwendung haben Bobaths diesen für das Verständnis und die Behandlung der Zerebrallähmung wesentlichen Annäherungsweg bereits erforscht.

Les reflexes de posture et de correction

De nombreux mécanismes posturaux et centres de coordination musculaire sont localisés dans la moelle épinière. Ce qui ne s'y trouve pas est un mécanisme provoquant le mouvement et le comportement moteur approprié à la totalité de l'influx sensoriel qu'il faut intégrer aux

besoins de la tête, porteuse des récepteurs de distance. Du fait du rôle important joué par l'oeil, non seulement pour la survivance de l'espèce, mais, chez l'homme, pour l'accomplissement de tâches visuelles spécialisées, la tête et le cou contiennent les organes sensoriels, centres collecteurs et distributeurs d'information, qui dominent et organisent les centres médullaires en activité modulée, amorcée, contrôlée, coordonnée et intégrée par l'influx afférent approprié.

Du fait de la large distribution, à l'intérieur du système nerveux, de centres contrôlant la posture, ainsi que de tractus d'interconnection, il n'est pas surprenant que spasticité, flaccidité, augmentation des réactions positives de renfort ou des réflexes toniques cervicaux, et autres désordres posturaux, soient aussi fréquemment rencontrés à l'occasion de n'importe quelle lésion du cerveau et de la moelle épinière. Cependant, une fois que le trouble postural est établi, il est nécessaire de l'étudier pour permettre une localisation approximative du niveau de la lésion et aussi de déterminer quelles manœuvres réflexes sont capables de le transformer en un mouvement prévisible, contrôlé et, par conséquent, utile.

Dans le domaine de l'application, les Bobaths ont déjà exploré cette voie d'approche fondamentale dans la compréhension et dans le traitement de l'infirmité motrice cérébrale.

REFERENCES

1. Bard, P. (1938) 'Studies in the cortical representation of somatic sensibility.' *Harvey Lect.*, **33**, 143-169.
2. Bobath, K. (1961a) 'The nature of the paresis in cerebral palsy.' 2nd N.S.S. International Study Group on Child Neurology and Cerebral Palsy. *Little Club Clin. dev. Med.* (In the Press).
3. — (1961b) 'The long term results of treatment.' *Ibid.*
4. Denny-Brown, D. (1929a) 'On the nature of postural reflexes.' *Proc. Roy. Soc. B*, **104**, 252-301.
5. — (1929b) 'The histological features of striped muscle in relation to its functional activity.' *Ibid.*, pp. 371-411.
6. — (1960) 'Motor mechanisms—the general principles of motor integration.' *Handbook of Physiology. Neurophysiology II*, chap. 32, 781-786. Washington, D.C.: American Physiological Society.
7. Granit, R. (1955) *Receptors and Sensory Perception*. New Haven: Yale University Press.
8. Liddell, E. G. T., Sherrington, C. S. (1924) 'Reflexes in response to stretch (myotatic reflexes).' *Proc. Roy. Soc. B*, **96**, 212-242.
9. — (1925) 'Further observations on myotatic reflexes.' *Ibid.*, **97**, 267-283.
10. Magnus, R. (1925) 'Animal posture.' *Ibid.*, **98**, 339-353.
11. — (1926) 'Some results of studies in the physiology of posture.' *Lancet*, **ii**, 531-536, 585-588.
12. Rademaker, G. G. J. (1930) 'Expériences sur la physiologie du cerveau.' *Rev. Neurol.*, **1**, 337-367.
13. Rushworth, G. (1958) 'Muscle sense organs and disorders of movement.' *Cerebral Palsy Bull.*, **1**, No 3, 1-5.
14. Sherrington, C. S. (1915) 'Postural activity of nerve and muscle.' *Brain*, **38**, 191-234.
15. — (1947) 'The integrative action of the nervous system.' London: Cambridge University Press.
16. von Tschermak, A. (1952) 'Introduction to physiological optics.' Translated by P. Boeder. Springfield: Thomas.
17. Whitteridge, D. (1960) *Handbook of Physiology. Neurophysiology II*, chap. 42, 1089-1109. Washington, D.C.: American Physiological Society.

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Disturbances of Manual Perception in Children with Cerebral Palsy

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THE difficulties encountered in the therapeutic training of children with cerebral palsy are not exclusively motor.

In addition, these children often have disorders of perception, sometimes called 'gnostic disorders', from a Greek word meaning knowledge or recognition. They cannot tell what position in space the various parts of their limbs occupy; they do not know whether their joints are flexed or extended. When their eyes are shut they cannot recognise objects put into their hands. The work of Piaget, Zazzo, J. P. M. Tizard, Richmond Paine, Bronson Crothers, Tournay, Stella Albitreccia, and Chemama has raised widespread interest in these disorders.

It is easy to imagine how these disorders might interfere with motor training, either by preventing relaxation techniques from reducing hypertonia or by preventing the children learning the complex actions which enable them to cope with everyday life. Gnostic and praxic disorders inevitably go together.

In some cases, the evaluation of gnostic disorders and their correction by re-education may be the first necessity. If these disorders are very pronounced, their treatment must precede that of the motor disorders.

The first question to ask in making such an evaluation is: on what grounds can one say that gnostic disorders exist? When the damage is unilateral one can, of course, compare the sound side with the hemiplegic side, and in adults, where the normal

gnostic possibilities are at least roughly known, the diagnosis of bilateral agnosia can be made without too much difficulty, if with little precision. On the other hand, in children with bilateral cerebral palsy it is essential to make a strict comparison

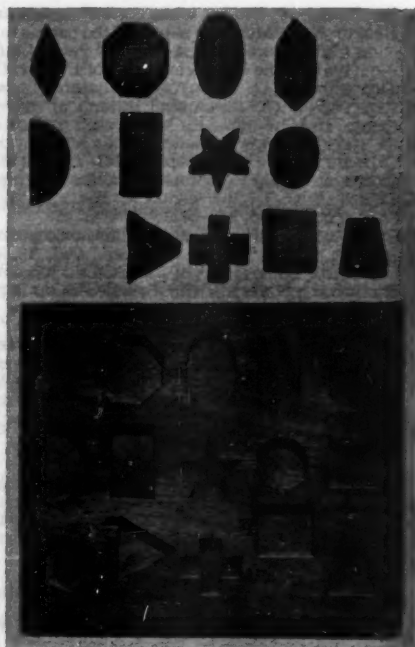


Fig. 1. Above: Geometrical shapes used for the study of manual perception in children aged 4-7 years. Below: Board used (with corresponding shapes) for the study of visual perception (Grace Arthur). The board measures 37 x 20 cm.

between their responses and those of normal children of comparable chronological and mental age.

Preliminary Study

A preliminary study of normal children was therefore made. This related only to the recognition of shapes held in the hand

by 218 normal children between the ages of 2½ and 8½ years.

The children were asked to recognise the following objects without looking at them. (1) A pencil, a marble, a cube, a box and a cotton-reel. (2) The twelve geometrical shapes illustrated in Fig. 1, always offered in the order shown on the

Right side				EXAMINATION SHEET	Left side			
Date	Date	Date	Date		Date	Date	Date	Date
				Actual age Mental age 'Gnostic' age				
				Cube Marble Pencil Cotton reel Box				
				Round Square Triangle Lozenge Hexagon Octagon Oval Semi-circle Star Cross Trapezium Rectangle				

Fig. 2. Examination sheet used for calculating the child's gnostic age, showing the difference between the right and left sides.

examination sheet in Fig. 2. These shapes were not manipulated by the children, but merely put into their hands by the tester. This precaution was taken because the disabled children examined later would be incapable of manipulating the test objects.

The children were not required to name the shape but to point it out among all the shapes when the cloth which hid them during manipulation was drawn away.

It was found that normal children of 2½ years (and even of 2 years) can recognise the ordinary objects without difficulty. At 4 years normal children recognise 5 to 7 of the geometrical shapes. At 5 years they recognise 7 to 8 shapes. At 6 years they recognise 9 shapes. At 7 years they recognise all the shapes and thereafter the test is no longer discriminative.

On the whole, the shapes most easily recognised are the circle and the star. The most 'difficult' shapes are the cross, the trapezium and the hexagon.

When these facts were known it became possible to find out whether manual perception was as well developed in a child with cerebral palsy as in a normal child of the same age.

Present Study

The present study relates to 92 children with cerebral palsy, a number large enough for statistical evaluation. It cannot, however, be taken to show the incidence of gnosic disorders in children with cerebral palsy, in general, since the children examined were not taken at random from those who came to us for consultation but were selected from 2,000 case-histories.

In fact, for practical reasons, the testing of manual perception was mostly done on inpatients of the Re-education Department of Garches and Bicêtre. This means that the children had already been selected, and had been judged, by their level of intelligence and emotional development, to be

capable of benefiting from re-education and capable of adapting themselves sufficiently to temporary separation from their families and to life in a community. We retained only those histories from which an adequate psychological evaluation could be made.

In some cases we confined ourselves to recording *N* (normal). This was done in children who either followed the normal school curriculum or had not been able to undergo complete psychological tests at the consultation, but who gave answers normal for children of their age when the test objects were put to them.

It is essential to know the children's general standard of intelligence. In order to define the gnosic disorders present, and hence to give advice on gnosic re-education, it is not enough to establish that the disabled child's manual perception is less well developed than that of a normal child of the same chronological age. Often, in fact, the child with cerebral palsy is deficient in all-round mental development. It is not surprising, then, that when one examines such a child, one may find that manual perception is not so well developed as in a normal child of the same age, but is equal to that of a child of the same mental age. Only when the 'gnosic age' is less than the mental age can one speak of a relative agnosia and advise gnosic re-education.

All the results which are abnormal according to this definition are shown in bold figures in the tables. The child's actual age is also shown in the tables, 'age (1)' being the child's actual age at the time of the psychological test, and 'age (2)' that at the time of the 'gnosic' test. Mental age is stated at the time of the psychological test.

Knowledge of the global mental age is not sufficient in itself to establish the presence of a *manual agnosia*. One must also know that shapes not recognised in

TABLE I—FINDINGS IN 92 CHILDREN WITH CEREBRAL PALSY OF THE TYPE INDICATED.

The bold figures indicate Severe Agnosias and the italics Mild Agnosias. 'Age (1)' is the child's age when the psychological test was done, and 'Age (2)' when the gnostic tests were done. (Ages in years and months).

Case No.	Sex	Side most affected	Psychological test	Age (1)	Mental age †	Age (2)	Gnostic age	
							Right	Left
Unilateral Spasticity								
1	M	R	Gesell	3	3	5:8	3	4
2	M	R	Terman	5:5	5	5:8	3	4
3	M	R	Terman	13:1	11:9	13:1	3	IV 6
4	F	R	—	—	N	14	IV 6	IV 6
5	F	R	Wisc	14:5	≥ 14:5	14:7	IV 3	6
6	M	R	—	—	N	12	IV 4	IV 6
7	F	R	Terman	9:5	9	9:5	4	IV 6
8	M	R	Terman	5:5	4:4	5	3	3
9	M	R	Columbia	9	6:2	9	IV 3	IV 6
10	M	L	Terman	9:5	6:10	9:5	IV 6	3
11	M	L	Terman	4:6	3:5	4:4	3	3
12	M	L	Wisc	6:5	5:11	6:5	4	3
Unilateral Rigidity								
13	M	R	Columbia	11:10	7:10	11:10	3	6
14	F	R	Terman	10	10	11	6	6
15	M	L	—	—	N	14	6	6
Unilateral Athetosis								
16	M	R	Terman	11:5	9:4	9:4	< 3	6
17	M	R	Columbia	5:8	4:5	7	6	6
18	M	R	Terman	8:2	2:3	7:10	3	6
19	M	R	Terman	14:8	6:7	14:8	4	4
20	M	R	Terman	6:8	4:6	7	6	6
21	F	L	Gesell	3	3	5:1	6	4
Unilateral Hypertonia								
22	M	L	Terman	9:6	8:2	9:10	IV 6	IV 6
Bilateral Spasticity								
23	M	—	Terman	8:10	7:2	9:11	IV 6	IV 6
24	M	R	Columbia	6:6	6:2	6:2	3	3
25	F	—	Terman	11:6	≥ 11:6	11:6	3	3
26	F	R	Terman	7:8	5:2	7:7	4	4
27	F	—	Terman	7:7	6	7:7	IV 6	IV 6
28	M	—	Matrix 1938	6:10	7:5	6:10	6	3
29	M	—	Terman	13	10:6	13	6	3
30	M	R	Terman	9	IV 6	9:9	IV 3	IV 6
31	F	R	Terman	9:1	4:11	9:1	3	4
Bilateral Rigidity								
32	M	R	Wisc	9:3	7:2	9:4	IV 6	IV 6
33	M	R	Terman	9:9	6	10:3	IV 3	6
34*	M	R	Terman	4:8	4:10	4:2	4	3
35	F	R	Terman	9:11	6:11	9:10	4	6
Bilateral Athetosis								
36	M	—	Terman	15	15	15	IV 6	IV 6
37	M	L	Terman	7:11	6	7:11	IV 6	IV 6
38	F	—	Impossible	—	—	7	4-5	6
39	M	R	Terman B.S.	10:5	7	10:9	IV 6	IV 6
40	M	—	Columbia	9:10	9:10	9:10	3	3
41	M	—	Columbia	7:10	4:9	8:5	3	3
42	F	—	Wisc	6:6	4:2	6:1	4	4
43	M	L	Terman	8:4	6	8:3	IV 6	IV 6

* This boy writes more readily with his right hand than his left, although his bilateral rigidity is more marked on the right side. † Estimated at the time of the psychological examination.

TABLE I—continued.

Case No.	Sex	Side most affected	Psychological test	Age (1)	Mental age †	Age (2)	Gnostic age	
							Right	Left
44	M	L	Terman	5:7	4:5	10:11	IV 6	4-5
45	F	R	Terman	3:11	4	4:6	3	4
46	M	—	Columbia	5:8	5:8	5:7	5	5
47	F	—	Wisc	9:5	7:8	9:3	5	5
48	M	R	Terman	4:8	4:9	4:8	IV 3	IV 3
49	M	—	Terman	8:8	6:10	8:8	IV 6	IV 6
50	M	R	Columbia	8:2	4:5	8:7	IV 6	4
51	M	—	Columbia	6:5	7:10	4:9	IV 6	4-5
52	F	L	Terman	7:7	6	7	IV 6	5
53	F	L	Terman	10:6	10	12:1	IV 6	IV 6
54	M	R	Terman	6:4	5:4	8:10	4	4
55	M	R	Terman	4:3	2:8	4	3	3
56	F	—	Terman	10:4	7:1	10:4	IV 6	IV 6
57	M	—	—	—	N	10	IV 6	IV 6
58	M	R	Terman	4:8	4:10	4:6	4	5
59	F	—	Terman	10	IV 8	10	IV 6	IV 6
60	F	—	Terman	14:4	7:6	14:4	4-5	3
61	F	—	Terman	8:11	5:1	8:11	4	5
62	M	—	Terman	13	8:8	13	IV 6	IV 6
63	M	R	Terman	9	6:6	8:10	IV 6	IV 6
Bilateral Hypertonia								
64	M	L	Wisc	9:2	6:5	9:10	IV 6	IV 6
65	M	—	Gesell	6:9	3-3:6	6:9	IV 3	IV 3
66	M	R	Terman	11	9	11	IV 4	IV 6
67	F	R	Columbia	9	8	9	IV 6	IV 6
68	M	L	Terman	6:1	6:8	6:6	4	5
69	M	—	Wisc	6:2	5:2	5	4	6
70	M	—	Terman	8:5	7:5	8:5	IV 6	5
71	M	—	Terman	4:9	4:4	4:7	3	3
72	M	L	Terman	8:8	6:10	8:8	IV 6	IV 6
73	F	—	Gesell	5:2	2	5:2	3	3
74	M	—	Terman	8	7	8	4	4
Ataxia								
75†	F	—	Wisc	8:3	6	8	4	6
76	F	—	Wisc	6:8	3:8	6:8	4	4
77	F	L	Terman	7:8	7:8	7:8	5	4
78	F	—	Columbia	7:4	7:5	7:5	IV 6	IV 6
79	F	—	Wisc	6:4	5:6	6:1	IV 6	IV 6
80	M	—	Gesell	4:1	3	4:1	3	3
81	M	—	Terman-Merrill	8:8	5	8:7	5	6
82	M	—	Binet-Simon	8:4	5	8:5	4	IV 6
83†	F	—	Terman	5:1	4:4	4:9	3	3

† Shows athetotic movements. ≥ means 'more than or equal'. ≤ means 'less than or equal'.

the hand can be recognised by sight. If the child fails in this test, one can no longer speak of a separate manual agnosia. One may then be dealing with a disordered appreciation of spatial relationships, either from lack of education (we have noted the influence of schooling on normal children), or simply from lack of concentration or attention on the part of the disabled child. We would draw attention to case 18,

where the child's gnostic age is far higher than his mental age.

Motor Disorders

The motor system was examined, as a routine, in all the cases included in this study.

Within the limits of this article we cannot define the exact meanings we assign to the terms 'spasticity', 'rigidity' and

'athetosis'; they have been defined elsewhere. It sometimes happens, however, that the nature of hypertonia is clinically debatable, or that in a single limb some groups of muscles are affected with a certain type of hypertonia and other groups with another type. In these cases we have been content to speak of 'hypertonia', without going into further detail.

Among the 9 cases of ataxia (Cases 75-83) there were 2 in which athetotic movements could be detected in addition. We have indicated this with 'daggers' in Table I.

In 9 cases the motor disorders were so much greater in the lower limbs that real disablement of the upper limbs could hardly be said to exist (Table II).

Summarising, among the 92 children examined we find:

- 22 *unilateral syndromes* (hemiplegias), comprising—
 12 spasticity (9 right, 3 left)
 3 rigidity (2 right, 1 left)
 6 athetosis (5 right, 1 left)
 1 'hypertonia' (left).
 61 *bilateral syndromes*, comprising—
 9 spasticity (4 predominantly right)
 4 rigidity (all predominantly right)
 28 athetosis (8 predom. right, 5 predom. left)
 11 'hypertonia' (2 predom. right, 3 predom. left)
 9 ataxia.
 9 with disability largely or wholly confined to lower limbs.

Findings in Detail

In the 12 cases of *unilateral spasticity*

(Cases 1-12), a disorder of manual perception was recorded in 6 cases on the affected side, and in 4 cases on both sides.

There are only 2 cases in which the gnosis age is not less than the mental age, on either the affected or the sound side. In Case 6, however, it appears that the boy forms a conception of the outside world in a very distinctive manner. On the sound side, he behaves like a normal child in arriving immediately at a synthesis. His responses are: 'It is a circle', or 'It is a half circle'. Sometimes he hesitates briefly: 'It isn't a square, it's a rectangle'. On the other side he has to go through a process of analysis to recognise the shape and often replies only after some hesitation. Here are his responses verbatim: 'I know that on one side it is pointed . . . it isn't round. It seems to me that all its sides are the same length. It is a triangle because there's a side missing.'

In the 3 cases of *unilateral rigidity* (13-15) a disorder of manual perception is present in only 1 case, and affects only the side with the rigidity.

Of the 6 cases of *unilateral athetosis* (16-21) relative agnosia exists in only 3 (in 2 cases on the affected side only and in the third on both sides).

If we add the case of *unilateral 'hypertonia'* (22), where there is no disorder of manual perception, we find that, of the 22

TABLE II—FINDINGS IN 9 CHILDREN WITH CEREBRAL PALSY MAINLY AFFECTING THE LOWER LIMBS.

Case No.	Sex	Disability in upper limbs	Side not affected	Disabilities in lower limbs	Psychological test	Age (1)	Mental age	Age (2)	Gnosis age	
									Right	Left
Cerebral Palsy mainly in Lower Limbs										
84	F	Slight hypertonia	L	Spasticity	Terman	9:9	7:8	9:10	≧6	≧6
85	M	—	—	Spasticity	Terman	5:7	2:11	5:7	3	3
86	M	Debility	—	Spasticity	Wisc	8:10	5:6	8:9	III 3	III 3
87	F	—	—	Spasticity	Terman	5:5	3:9	5	3	3
88	F	Debility	—	Spasticity	Terman	5:7	2:11	5:7	3	3
89	M	Tremor	—	Spasticity	Terman	9:6	9:1	9:6	6	IV 6
90	M	—	—	Hypertonia	Terman	8:2	6	9:5	IV 6	IV 6
91	F	—	—	Spasticity	Terman	9:5	6:9	9:5	5	6
92	M	—	—	Athetosis	Terman	9:2	7:4	9:4	IV 6	IV 6

'hemiplegics', 5 have bilateral and 9 unilateral disorders of perception. The side of the hemiplegia apparently does not affect the frequency of the gnostic disorders: of the 8 cases without any disorder of perception, 6 came from the group of 16 right-sided hemiplegics and 2 from the 6 left-sided hemiplegics.

It is worth noting that bilateral gnostic disorders may be present when the motor damage is unilateral. Indeed, a hemiplegic child's sound side often shows some motor disability, such as a slight 'paratonia'* when he tries to touch his fingers one after the other quickly with his thumb. This is a negligible disability compared to the genuine disorder on the opposite side.

The gnostic disorders can affect the two hands indiscriminately. Here again the side affected by the motor disability does

* Paratonia (Dupré 1907) signifies an excessive use of muscles in the performance of a task compared with that shown by another subject of the same mental age.

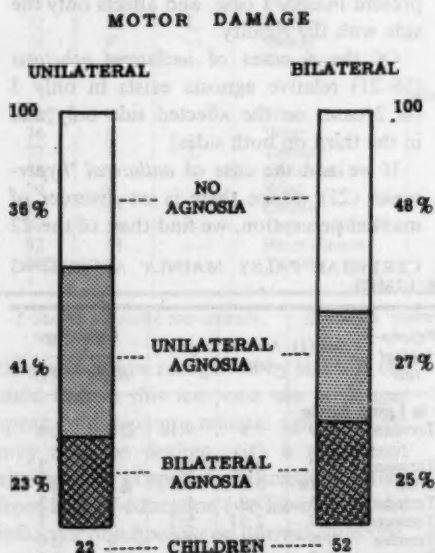


Fig. 3. The gnostic disturbance may be bilateral when the motor damage is unilateral, and conversely.

not seem to make any difference: of the 5 cases with bilateral gnostic disorders, 4 came from the 17 right-sided hemiplegics, and 1 from the 5 left-sided hemiplegics.

A different result is obtained when the cases are classified according to severity. It can be assumed that severe cases are

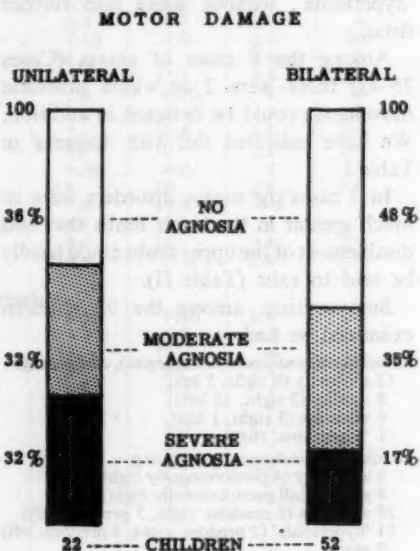


Fig. 4. Gnostic disturbances are probably commoner and more severe in unilateral than in bilateral cases.

those with a gnostic quotient of under 50, the gnostic quotient being: $\text{gnostic age} \div \text{mental age} \times 100$. Of the 7 severe cases revealed in this way no less than 6 affect the right side.

Of the 9 cases of *bilateral spasticity* (23-31), all but 2 have gnostic disorders. In 3 cases these disorders are bilateral, and in 4 cases unilateral. In at least 2 of these last 4 cases the disorders affect the limb with the most motor damage.

Of the 4 cases of *bilateral rigidity* (32-35), 1 has no gnostic disorders. In the other 3 cases a gnostic defect is present on one side. One of these cases (34) is very

instructive. The boy's motor disorders are more marked on the right side, the gnostic defect is present only on the left, and the child prefers to use his right hand for writing but his left for other tasks.

Of the 28 cases of *bilateral athetosis* (36-63), 18 have no gnostic disorders, including Case 39, where the boy has such marked motor disorders in his upper limbs that he cannot use them at all. Bilateral gnostic disorders are present in 6 of these cases and unilateral ones in 4 cases. In at least 3 of these cases it is the limb most affected from the motor point of view that has also suffered most from the gnostic aspect.

Finally, among the 11 cases of *bilateral 'hypertonia'* (64-74) there are: 4 cases with no gnostic disorders; 4 with bilateral gnostic disorders; and 3 with unilateral gnostic disorders.

If we compare the frequency and severity of the gnostic disorders in the 22 'hemiplegics' with those in the 52 children with bilateral motor disorders, we find no

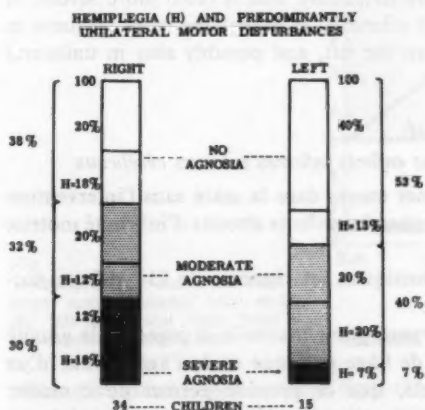


Fig. 5. Possible influence of the side of the motor damage on gnostic disturbance. When all cases of agnosia are considered, the side of the damage probably makes no difference, but severe agnosia seems more common when the motor damage is right-sided.

significant differences* (Figs. 3 and 4).

Bilateral agnosias: 5/22 against 13/52 ($\chi^2 = 0.04$).

Unilateral agnosias: 9/22 against 14/52 ($\chi^2 = 1.35$).

Severe agnosias: 7/22 against 9/52 ($\chi^2 = 1.8$).

No agnosia: 8/22 against 25/52 ($\chi^2 = 0.9$).

In bilateral cerebral palsy, it is possible that the side with the greater motor

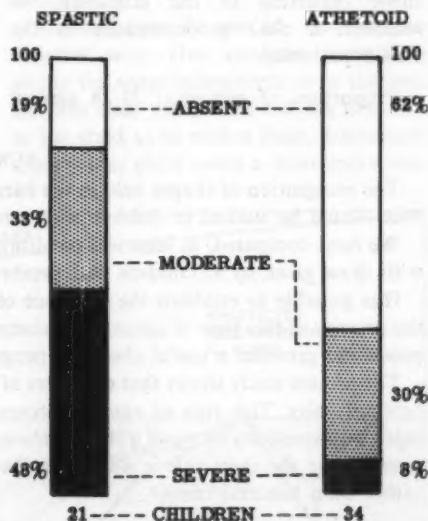


Fig. 6. Gnostic disturbances are commoner in spastic than in athetoid children, whether all cases of agnosia or only the severe cases are considered; the difference is statistically highly significant.

disorder may have an influence on the frequency and severity of the gnostic disorders, but our figures are not large enough for the differences to be significant. Of the 18 predominantly right-sided cases, there are only 7 without agnosia, while of the 9 predominantly left-sided cases, there are 6 without agnosia; χ^2 , however, is only 1.8.

* The difference would be significant at the level $P = 0.05$ if $\chi^2 > 3.84$.

Among the 18 predominantly right-sided cases there are 4 with severe agnosia. Of the 9 predominantly left-sided cases there are none with severe agnosia, but this would only be significant at the level $P = 0.17$, which is insufficient (Fig. 5).

If we now take the unilateral and bilateral types as a whole, and compare the frequency, and above all the severity of the gnosic disorders occurring in the 'true' hypertonias (spasticity and rigidity) with those occurring in the athetoses, we establish a clear predominance in the true hypertonias.

Proportion of agnosias: 21/28 against

13/34 ($\chi^2 = 2.32$, not quite significant).

Proportion of severe agnosias: 12/28 against 3/34 ($\chi^2 = 13.5$, highly significant).

In the cases with spasticity the results are even clearer, since out of 21 cases there are 17 agnosias, 10 of them severe (Fig. 6).

Among the *ataxias* (75-83), disorders of manual perception are again common (4 cases out of 9), but no severe agnosia can be found.

It is strange to record that we also found agnosia in 2 of the 9 cases where the motor disorders were clearly confined to the lower limbs (Table II).

SUMMARY

The recognition of shapes held in the hand without the help of sight is one of the gnosias that should be studied in children with cerebral palsy.

We have compared, in identical conditions, the responses given by 218 normal children with those given by 92 children with cerebral palsy.

It is possible to establish the existence of disorders of manual perception, and to judge the severity of this type of agnosia, by comparing a child's gnosic age and mental age. This procedure provides a useful check on progress during re-education.

The present study shows that disorders of manual perception can be found in all forms of cerebral palsy. This type of agnosia occurs more frequently, and is often more severe, in cases with spasticity or rigidity than in those with athetosis. It is perhaps more frequent in cases where the right side is affected rather than the left, and possibly also in unilateral rather than bilateral cases.

RÉSUMÉ

Troubles de la perception manuelle chez les enfants infirmes moteurs cérébraux

La faculté permettant de reconnaître les formes tenues dans la main sans l'intervention visuelle est une des gnosies que l'on doit étudier chez les enfants atteints d'infirmité motrice cérébrale.

Nous avons comparé, dans des conditions identiques, les réponses de 218 enfants normaux et celles de 92 enfants infirmes cérébraux.

On peut fonder l'existence de troubles de la perception manuelle et juger de la gravité de ce type d'agnosie sur la seule comparaison de l'âge gnosique et de l'âge mental d'un enfant. Nous verrons, dans le prochain article, que ce procédé permet de contrôler utilement le progrès survenant au cours de la ré-éducation.

La présente étude prouve que les troubles de la perception manuelle peuvent se rencontrer dans toutes les formes d'infirmités motrices cérébrales. Ce type d'agnosie est plus fréquent, et souvent plus grave, dans les cas avec spasticité et rigidité que dans ceux avec athétose. Peut être le rencontre-t-on plus souvent dans les cas où le côté droit est plus atteint que le gauche et aussi dans les cas de paralysie unilatérale plutôt que bilatérale.

Development of Manual Perception in the Child with Cerebral Palsy during Re-Education

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In our previous article in this *Bulletin* we showed how a child's gnostic age could be established with reference to the recognition of shapes held in the hand. We then described in detail how to establish the existence and severity of agnosia in the child with cerebral palsy. We agreed that a relative agnosia was present in all cases

how these relative agnosias may come about.

Re-education is carried out in the simplest way. One confines oneself to giving the same instructions as in the test, but this time one points out his mistakes to the child as he makes them. Sometimes a particular child needs a more individual

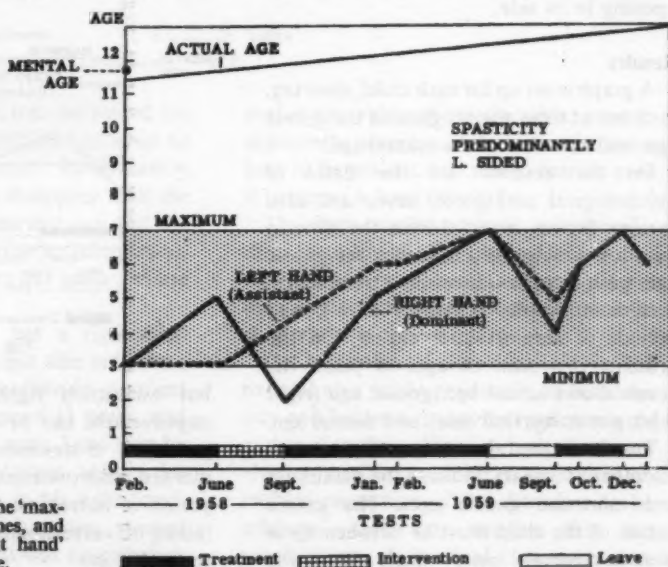


Fig. 1.
Note that in these charts the 'maximum' and 'minimum' lines, and the 'left hand' and 'right hand' curves refer to gnostic age.

where the gnostic age was less than the mental age, and that this agnosia was severe when the gnostic age was less than half the mental age.

We should like to show here how one can follow the evolution of these disorders during re-education, and to ask ourselves

technique. For example, unreliable children with poor powers of concentration have least difficulty if, to begin with, they are asked to choose between only three or four shapes. Subsequently the number is progressively increased.

The child sometimes finds it easier to

recognise blocks which are either larger or taller than the shapes used in the test, such as the Kiddicraft letter blocks.

In some hemiplegics one may confine oneself at first to putting a shape such as a square into the affected hand. Later all the various shapes are put first into the affected hand and then into the sound one, and from this passing process the child has to learn to recognise that he has held the same shape in each hand. Sometimes one gets better results by making the child analyse what he feels than by asking him to recognise the shape as a whole (whether it is flat, rounded, pointed, etc.).

It is worth while asking the child to recognise tools without looking at them by putting his hand into a toolbox through an opening in its side.

Results

A graph is set up for each child, showing, in terms of time, the progress of the gnosic age, and also that of the mental age.

On the abscissa are the dates of psychological and gnosic tests, and also various factors noted during the time in question which could influence the gnosic age: periods of re-education ('treatment'), periods of family holiday ('leave'), and periods of surgical intervention. On the ordinate is a scale of ages in years, the graph shows actual age, gnosic age (right side), gnosic age (left side), and mental age.

Two horizontal lines, one at 7 years and the other at 3 years, indicate the maximum and minimum gnosic ages. The gnosic curves of the child must lie between these two horizontal lines, which form the agreed limits of our test. At present we do not know how to investigate manual perception in children under 2 years of age or over 6-7 years.

In some cases the influence of re-education is evident in the graphs.

The child in Fig. 1, who has bilateral spasticity with clear left-sided pre-

dominance, has severe gnosic disorders, as the 'gnosic quotient' is only 25 to begin with on both the right and the left sides (gnosic age 3, actual age 11½ years; I.Q. 100). At the outset progress is more evident on the right than on the left, and then, after surgical intervention on the lower limbs, there is a relapse on the right side, while progress on the left side continues. Periods of leave involve some falling off, and this shows well the efficacy of re-education.

Next we have a similar graph for the child in Fig. 2, who has an equally severe

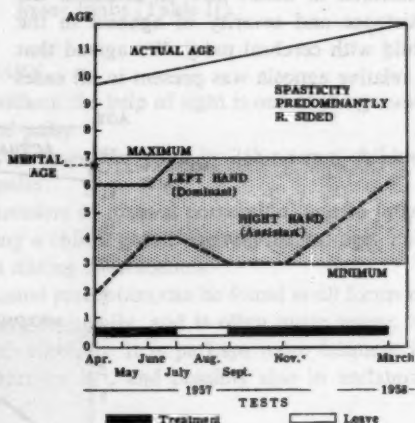


Fig. 2.

but exclusively right-sided agnosia. An improvement can be seen during the first period of in-treatment, and a still more marked improvement during the second period of in-treatment, whereas a definite falling off corresponds to the child's return to his family.

In the case of the child in Fig. 3, progress has been slower. This child with bilateral but predominantly right-sided rigidity, had, to begin with, gnosic disorders which were only moderate and confined to the left side. Whereas no progress at all was noted on the right side, manual perception became as good in the left hand as in the

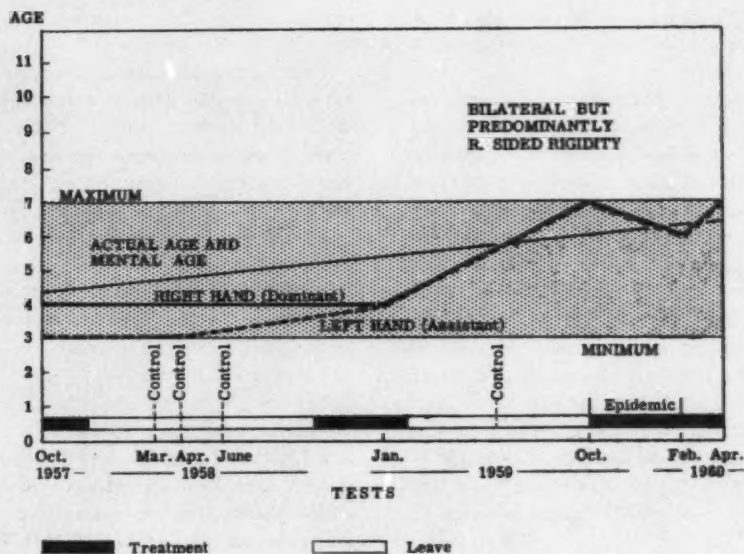


Fig. 3.

right, but that of both was backward for the child's mental age (which was equal to his actual age in years). Re-education caused the disorders to disappear until the gnosis age passed the mental age and the actual age as well. During an intercurrent illness there was a short but quite definite falling off.

The boy in Fig. 4 has a right-sided hemiathetosis. On the right side only there is considerable agnosia (gnostic quotient less than 20). At the age of 11 years, and with a mental age of 9 years 3 months, he could not recognise common objects, let alone geometric shapes, in his right hand. For 18 months, re-education seemed doomed to failure, but it was nevertheless continued. Since that time the boy has made regular progress, with only one period of marking time during a stay with his family.

Discussion

From these few examples the first inference to be drawn is that one should

realise the value of re-education, even in apparently severe cases. One must know how to be patient, and not to despair even if progress is long delayed.

Do these results warrant any speculations from the physiopathological point of view? Two possible hypotheses present themselves to explain relative agnosia in children with cerebral palsy:

(1) A cerebral lesion localised in the parietal area might produce it, in the same way as a lesion in the motor tract produces motor disorders. Since substitutions are always possible in children, re-education enables correction of the disorders to be achieved. One can imagine that the possibilities of gradual learning may depend on the extent of the zones which can be used as substitutes, and in particular on the extent of the association areas. This is why, in our opinion, hope is justified only when there is a clear discrepancy between the gnosis age and the mental age. This difference, especially when it is estimated by tests highly loaded in

"g" factor, seems to us to allow the value of the association areas to be assessed to some extent.

(2) Another hypothesis is often put forward. It is right to repeat, particularly in articles and books meant to be read by the families of these children, that because of his motor disorders the child with cerebral palsy cannot take possession of the objects which surround him. He can know them only by sight. It is therefore reasonable to suggest that this absence of prehension entails disorders of manual perception and of body image. Therefore one can only subscribe to the advice always given, that thesegnosias should be developed from the earliest possible age by passively moving the limbs, and by putting things into the child's hands as often as possible.

However, there are some confusing cases. For example, one child has such bad athetosis of the upper limbs that they are of no use to him whatever, but despite the

severity of the motor damage no gnosic disorder at all is present.

There are cases in which the theory that agnosia is caused not by a lesion but by lack of use seems reasonable. These are the cases in which the gnosic age rises quickly and overtakes the mental age. Obviously, one can always wonder whether the child has understood the test instructions properly, and if he has really paid attention. But when such success is obtained in cases where the gnosic disorders were both severe and strictly unilateral, the doubt seems hardly permissible. Thus, a boy aged 13½ years has a mental age of 10 years 9 months in performance tests, and 13 years 2 months in verbal tests. His gnosic disorders are strictly unilateral, which shows that he must have understood the instructions properly. In 15 days he rose from a standard of less than 3 years to one of 5 years. Three months later he had reached the ceiling of our test (a standard equal to or above 7 years).

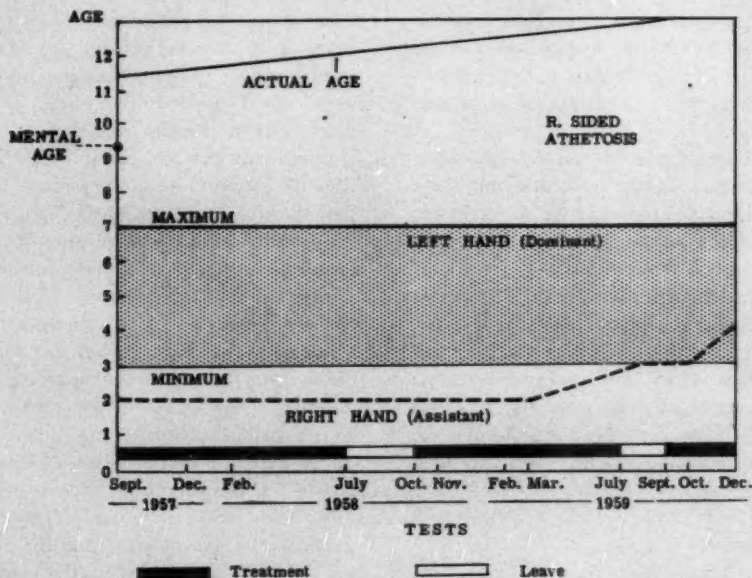


Fig. 4.

SUMMARY

In every case of cerebral palsy it is essential to examine the gnosias: recognition of objects held in the hand, recognition of the position occupied by the parts of a limb in space, tactile discrimination, body image, and the 'extinction phenomenon'. In the present study we have reported only on the progress made during re-education in the ability to recognise shapes held in the hand.

When we speak of the gnostic quotient we mean the relation between the gnostic age (established by comparison with the responses of children of the same mental age) and the mental age. Even when the gnostic disorders are profound (gnostic quotient less than 50), re-education can result in a considerable improvement, or in the disappearance of the disorders. This re-education sometimes has to be very prolonged and must not be interrupted. Fallings off are frequent when re-education is interrupted.

It seems permissible to contrast agnosias caused by a lesion, in which re-education produces only long-term success, with agnosias caused by lack of use, in which results are much more quickly obtained.

RÉSUMÉ

Développement de la perception manuelle chez l'enfant atteint de paralysie cérébrale pendant la rééducation

Dans tous les cas de paralysie cérébrale, il est essentiel d'examiner les gnosies: reconnaissance des objets tenus dans la main, reconnaissance de la position occupée par les différentes parties d'un membre dans l'espace, discrimination tactile, schéma corporel et 'phénomène d'extinction'. Dans la présente étude, nous avons seulement rapporté les progrès faits pendant la rééducation en ce qui concerne l'aptitude à reconnaître des formes tenues dans la main.

Quand nous parlons de quotients gnosiques, nous entendons le rapport entre l'âge gnosique (établi par comparaison avec les réponses d'enfants du même âge mental) et l'âge mental. Même quand les troubles gnosiques sont profonds (quotient gnosique inférieur à 50), la rééducation peut amener une amélioration considérable ou même la disparition des troubles. Cette rééducation doit être parfois très prolongée et ne doit pas être interrompue. Des rechutes sont fréquentes quand la rééducation est interrompue.

On peut opposer les agnosies causées par une lésion dans lesquelles la rééducation n'apporte une amélioration qu'à longue échéance et les agnosies causées seulement par absence d'utilisation dans lesquelles les résultats sont obtenus beaucoup plus vite.

ZUSAMMENFASSUNG

Entwicklung des Wahrnehmungsvermögens der Hand beim Kinde mit Zerebrallähmung während der speziellen Erziehung

In allen Fällen von Zerebrallähmung ist es wesentlich, die Gnosien zu untersuchen: Erkennen der in der Hand gehaltenen Gegenstände, Erkennen der Lage der verschiedenen Teile eines Gliedes im Raum, taktiles Unterscheidungsvermögen, Körperbild, und 'Auslöschungsphänomen'. In dieser Arbeit haben wir nur über die Fortschritte, die während der speziellen Erziehung im Bereiche der Fähigkeit, in der Hand gehaltene Formen zu erkennen, gemacht wurden.

Wenn wir über den gnostischen Quotient sprechen, so meinen wir das Verhältnis zwischen dem gnostischen Alter (durch Vergleich mit den Antworten von Kindern desselben Intelligenzalters festgesetzt) und dem Intelligenzalter. Sogar wenn die gnostischen Störungen

tief sind (gnosischer Quotient unter 50), kann die Erziehung eine erhebliche Besserung oder das Verschwinden der Störungen. Zur Folge haben. Diese spezielle Erziehung muss manchmal sehr lange fortgesetzt werden und man darf sie nicht unterbrechen. Rückfälle sind häufig wenn die Erziehung unterbrochen wird.

Es scheint möglich, die durch Läsion entstandenen Agnosien, in denen spezielle Erziehung nur nach langer Zeit erfolgreich ist, den durch Gebrauchsmangel entstandenen Agnosien gegenüberzustellen, bei denen die Resultate viel schneller erreicht werden.

Note: The material in these two papers by Dr. Tardieu and his colleagues formed the basis of a talk by Dr. J. C. Tabary at the N.S.S. Study Group on Hemiplegic Cerebral Palsy in Children and Adults in Bristol in September, 1961.

Pathological Anatomy of Anoxia

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ANOXIA implies a deficiency in oxygen supply to the tissues, and is a symptom and not a disease entity. It occurs in some degree and as a terminal event in all diseases. Anoxic conditions produce lesions which vary in different organs and with the duration as well as the severity of the anoxia. Few of the lesions are in any way specific for anoxic damage. Anoxic injury may determine death or may contribute to it, or it may allow survival with irreversible damage persisting as functional and structural deficiencies throughout life.

Anoxic lesions are often secondary and are then the result of abnormalities, injuries, infections and other diseases intrinsic to the subject. Anoxia is a special hazard of the process of adaptation to extra-uterine respiration, and intrinsic defects or deficiencies, inborn or acquired by the infant, are especially important at this period, but may also dictate the development of anoxic injuries at other periods of life. Anoxia arising from the environment, that is extrinsic to the individual and independent of him, is a special hazard of intra-uterine life, of birth and of the process of adaptation to extra-uterine life. This extrinsic (environmental) anoxia is responsible for just over half our intra-partum stillbirths and nearly half our deaths in the first three days of life. At other times it arises only from a few unusual accidents which include some complications of anaesthesia.

It is not enough to recognise that certain anatomical changes in the dead infant

have resulted from anoxia. It is necessary to decide if there is an intrinsic basis for the anoxia, and, if there is, interest must centre on that inborn or acquired defect, and not on the secondary and often terminal anoxia. If changes of anoxic damage are found, but no intrinsic cause exists, the pathologist must consider an extrinsic (environmental) basis. Then his task today is to probe the environment, and in the foetus or a newborn infant this means the maternal, obstetrical and placental environment, for the cause of the anoxia. Here, as in all fields of medicine today, the study of the pathological anatomy must be combined with every other approach, and, indeed, only when the cause resides in the placenta can anatomical study alone sometimes provide the basis. It may not be possible to recognise the basis of extrinsic anoxia, but 'anoxia' alone must never be regarded as a satisfying or final explanation. However, this important aspect of the pathology of anoxia cannot be further discussed now (see Table and Morison 1952).

ANOXIA

INTRINSIC (Secondary)	EXTRINSIC (Environmental)
Congenital	Maternal
Traumatic	Obstetrical
Inflammatory	Placental:—
Miscellaneous	Senescence
	Abruptio
	Infarcts
	Cord, etc.

Many conditions modify the anatomical expression of anoxia. In the perinatal period, when anoxia attains its greatest importance, both as a cause of immediate death and of postnatal injury, it is modified by the process of adaptation to extra-uterine respiration, by the maturity of the infant and by the duration of anoxia. As prematurity increases anoxic damage becomes an increasingly important cause of death. Sometimes, and usually after the neonatal period, an episode of anoxia dictated by some accident may result in cerebral devastation which is apparent even on gross study (Fig. 1). In other cases,

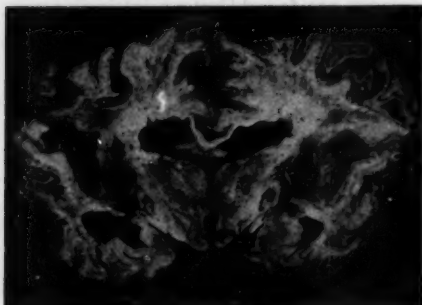


Fig. 1.
An anaesthetic accident at 14 days resulted in death at 98 days. The brain shows gross cortical and central atrophy.

an episode which is only slightly less severe may be responsible for brain damage which in post mortem material cannot be recognised or whose recognition may be debatable.

Some anatomical lesions of anoxia are general and affect the interstitial or supporting tissue of all organs, in varying degree and expression in the different tissues. Other lesions are more organ specific depending on changes in the special cells of the organ; their recognition may be more difficult. All the lesions of anoxia are most frequently and clearly seen in foetuses and newborn infants.

Anoxic Lesions Affecting Tissues Generally

Anoxia produces congestive circulatory failure with over-filling of the heart chambers and pooling of the blood in the viscera. In premature infants the vessels are thin-walled and poorly supported and become especially prominent and engorged. From the circulatory stasis and the anoxic injury to the endothelium petechial haemorrhages may result and these may dominate the picture, especially in premature infants. This is especially so if the intra-uterine anoxia has resulted from a large retroplacental haemorrhage, because this increases the pressure in the intervillous space, drives blood from the channels in the placenta and overloads the foetal circulation (Fig. 2). When this general tendency to haemorrhage affects certain organs, especially the brain, lungs and adrenals, it assumes a special importance.

Oedema of the tissues is another general manifestation of anoxia. It is little in evidence in anoxia of short duration, but, especially in continued anoxia occurring during intra-uterine life, it may be severe. A combination of vascular stasis and low oxygen causes the capillaries to leak fluid into the supporting tissues. Fluid escaping from the foetal blood in the foetal capillaries can be readily replaced from across the placenta from the much greater fluid reserves of the mother. After birth oedema fluid accumulated in the interstitial tissues may continue seriously to depress metabolic transfer between the cells in various organs and the circulating blood, to impair lung aeration and to impede free lung movement. These general disturbances, congestion, stasis, haemorrhage and oedema all in varying degree determine the lesions in the viscera, but changes in some parenchymatous cells also occur and may sometimes be recognisable.

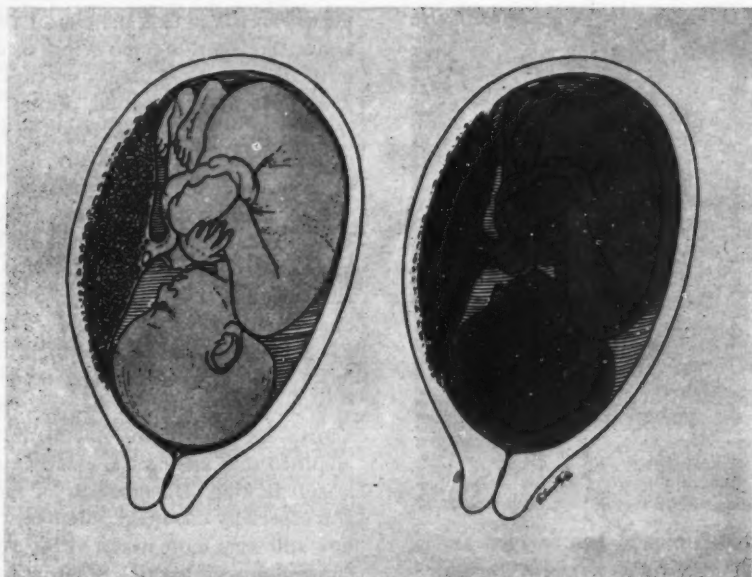


Fig. 2.

A retroplacental haemorrhage drives blood into the foetus from its placenta and overloads its circulation. Petechial haemorrhages in brain and viscera are often more prominent than in anoxia alone.

Anoxic Lesions in the Lung

The basic lesions in the lungs are congestion, the accumulation of oedema fluid in the air spaces and in loose tissues around the larger air passages and around blood vessels, and small but sometimes confluent haemorrhages into the air spaces. These are all non-specific lesions and appear in many disease conditions. Capillaries in the walls of the air spaces depend for their oxygen supply on the inspired air. As fluid accumulates in the air space system they are deprived of this and from their damaged walls more fluid escapes. In the words of Drinker (1945); 'Anoxia begets anoxia'. Oedema may thus be progressive but, since it may also accumulate rapidly, it is difficult to decide at post mortem whether any oedema present has contributed to death or whether it

has appeared only as an agonal event. Pulmonary oedema induced by anoxia at birth may progress and determine death hours or even days later, but this is difficult to establish.

In the foetus and newborn infant anoxia arising in intra-uterine life may produce other changes in the lung. Anoxic conditions during intra-uterine life can stimulate gasping inspirations which can draw amniotic fluid and the debris floating in it into the lungs (Fig. 3). Amniotic sac debris presents a distinctive histological appearance in the lung, but anoxia may develop after the liquor has drained away and may be fatal, when no amniotic material can be drawn into the lungs. Significant amounts of debris in the lung air spaces in the foetus or in the newborn mean that at some time the foetus has been exposed to anoxia. Its absence does not

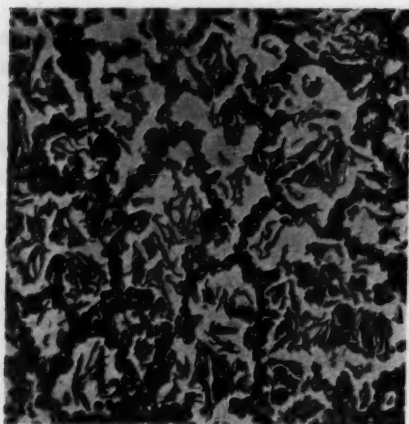


Fig. 3.

Inspiratory gasps dictated by anoxia draw vernix debris from the amnion into the lungs of the foetus.

mean that the child has escaped anoxic damage.

Anoxia causes the expulsion of meconium from the bowel and when this escapes into the amnion it may be inhaled into and may apparently block the larger air passages. However, many of these infants have suffered such severe anoxic damage that this mechanical blockage is of little significance, and most examples are seen in children dying during delivery.

There does appear to be a very close relationship between anoxic conditions before, during or immediately after birth and the development of hyaline membranes which may form in the first few days of life in the pulmonary air spaces. These membranes form in the alveolar ducts and close off the smaller air spaces where respiratory exchange occurs (Fig. 4). The mechanism of their production remains obscure, but their close connection with preceding anoxic conditions suggests that they are one way in which anoxic damage in the newborn progresses, often with remissions, to cause death hours or even one or two days later.

The central problem in the pathology of

the newborn is, indeed, this question of how anoxic damage sustained before or during birth dictates death hours or days later, and often after a period during which respiration has become established and anoxic conditions apparently abolished. Complicating congenital defects, intracranial and other injuries, infections, known anoxic lesions such as haemorrhage into the ventricles of the brain or into the adrenals and the formation of hyaline membranes must be excluded, but the mechanism by which anoxia itself dictates death often remains obscure. Some attribute these deaths to the delayed effects of anoxia on the brain, but it must be admitted that anatomical evidence of this is still lacking. Disturbances in the lung and especially disturbed pulmonary blood flow still seem more likely. Gross capillary congestion with haemorrhages into the air spaces is relative and non-specific. It is seen especially in premature infants in the syndrome of congestive pulmonary failure,

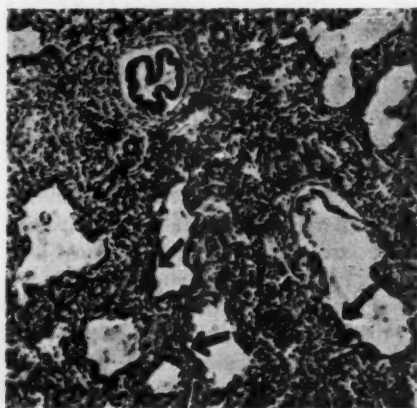


Fig. 4.

Hyaline membranes lining the smaller air passages prevent air entering air spaces which then collapse.

where the Uruguayan workers (Obes Polleri, Hill and Matteo, 1959) have very properly emphasised the overloading of

the pulmonary vascular bed. They consider that the persistent patency of the ductus arteriosus with the possibility of a reverse shunt from aorta to pulmonary artery is significant in the first few days after birth. This they have very ingeniously studied and illustrated by an electric circuit. The capillary bed of the lung must normally be protected by the lung arteries and arterioles, and it will be necessary to investigate the mechanism by which the blood vessels in the air-space walls are protected in the rapidly altering conditions imposed by the immediately postnatal circulation. Pathological anatomy must seek for deficiencies in the vessels of the lung which may lead to progressive pulmonary congestion and oedema and which result from anoxic damage before or during birth.

Anoxic Lesions in the Brain

The production by anoxia of intraventricular and sub-arachnoid haemorrhage, especially in the premature and newborn infant, is now fairly generally accepted. It is an important cause of early neonatal death. Great difficulties are experienced with the assessment of the significance of other interstitial changes, such as increased permeability of vessels and small capillary haemorrhages. Parenchymatous changes involving nerve cells present a special difficulty, and cytoplasmic changes of anoxia which may be recognised in the experimental animal cannot be recognised in human autopsy material where there must always be delay in fixation of tissue.

It is most essential that intraventricular haemorrhages arising in the walls of the lateral ventricles, and resulting in the escape of blood into the ventricular system and then into the subarachnoid space, be clearly distinguished from those haemorrhages which arise in the subdural space (Fig. 5). In the latter blood accumulates outside the arachnoid and comes from traumatic tears

of the great cerebral veins, veins lying in dural folds or from bridging veins. Failure to make this distinction and badly conducted autopsies invalidates many surveys even today. The incidence of intraventricular haemorrhages increases with prematurity. They are not related to the

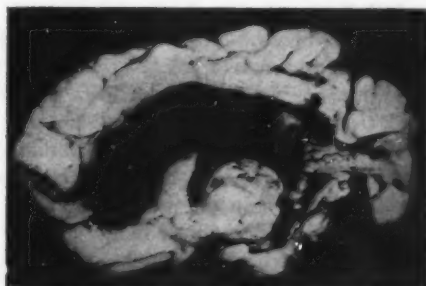


Fig. 5.
The blood clot in the lateral ventricle and the smaller haemorrhages deep to the ependyma are both due to anoxia.

difficulty of labour and are not due to physical trauma. It may be urged that sometimes strong intra-uterine contractions drive blood into the foetal head and over-distend and rupture the thin-walled and poorly supported vessels in the walls of the lateral ventricles. These lesions, however, occur after breech deliveries and elective caesarean sections, and the one constant feature is foetal, and often continuing postnatal, anoxia. They differ from other small haemorrhages, due to anoxia and present elsewhere in the body, only in the ease with which bleeding may occur into a fluid filled space.

It is more difficult to assess the significance of small haemorrhages often found on microscopical study of the brain, or to recognise and assess the significance of any increase or alteration in the fluid which may escape from the intracerebral blood vessels. Petechial intracerebral haemorrhages in the area of the respiratory centre and elsewhere do occur, but could easily be agonal and

be a terminal result and not the cause of anoxia. They are a theoretical rather than an observed cause of respiratory insufficiency or neonatal death after an anoxic delivery. It is doubtful if the excess of fluid escaping through the intracerebral blood vessels will cause oedema of the brain substance, or if its addition to the cerebrospinal fluid will disturb the intracranial pressure. Many histologists are too ready to accept various technical artefacts as evidence of brain oedema. In anoxia, and especially in premature animals, an increased permeability of the blood brain-barrier for dyes has been shown by Grontoft (1954). In the premature infant and probably in the anoxic infant at term the blood-brain barrier is unduly permeable to bilirubin and probably to potentially harmful substances.

For technical reasons routine sections of the neonatal brain are less satisfactory than those of older infants; maturity influences the normal distribution of nerve cells; migrating neuroblastic cells and occasional fat-laden macrophages are normal but are liable to be mistaken for the picture of pathological reaction. Nevertheless, the evolution of small cystic areas, most often in the deep white matter in the drainage area of the terminal veins and related to small anoxic haemorrhages often found there, has been traced by some workers, and the appearance in weeks or months of fibrillary gliosis and later of loose neuroglial scars noted. Cortical atrophy has also been related to a history of anoxia at birth (literature in Greenfield *et al.*, 1958). However, an assessment of the importance of neonatal anoxia in the aetiology of neurological disorders in later life must still depend on careful assessment of the evidence of anoxia. Even at autopsy there are no entirely specific changes which can be accepted as evidence of previous anoxic damage, and many of the late lesions of anoxia, of venous thrombosis and even of

kernicterus, not to mention congenital disturbances, may be indistinguishable. Recognisable lesions of anoxia are rarely appreciated in the neonatal period and their evolution into later childhood has been insufficiently studied. Apart from some cases of gross damage we still know far too little of the nature, incidence and significance of anatomical changes in anoxic brain damage. Few realise the amount of detailed work necessary for proper studies on cerebral anoxia.

Other Anoxic Lesions

In other organs haemorrhages tend to dominate the picture. Anoxic lesions in the adrenals are rare; they present as bilateral confluent haemorrhages in the cortices simulating those of birth trauma. Haemorrhages into the mucosa of the oesophagus, stomach and intestine may sometimes be important. Parenchymatous lesions such as focal hydropic vacuolation of liver cells and small focal necroses in the myocardium or in the media of arteries are rare.

It is more important to emphasise here that extrinsic anoxia increases greatly the risk of death from infection, and also increases the risk of bleeding from traumatic injuries. It facilitates lung infection by depressing respiratory reflexes, and in the oedematous and imperfectly aerated lungs organisms gain entry and multiply unchecked by cellular defences. Sometimes infection also spreads in connective tissues made oedematous by anoxia. Small amounts of blood within the subdural space often indicate only trivial birth trauma which has been associated with an anoxia arising independently which has facilitated bleeding from over-distended channels. Death often results from the anoxia and not from the presence of the blood, but in some cases it may be difficult to decide if anoxia or traumatic haemorrhage is the cause.

SUMMARY

Anoxic lesions may be secondary to intrinsic defects within the individual, and only when they result from environmental conditions and are extrinsic should they, rather than the initial defect, be recognised as a basis for death. Though of great importance, the primary maternal, placental and obstetrical basis for extrinsic anoxia in the foetal and neonatal period is not further discussed here.

The predominant lesions of anoxia result from congestion of vascular channels, haemorrhages from small blood vessels and the escape of oedema fluid. The lesions vary somewhat in the different organs. In the lungs the importance of the control of blood flow through the lungs at birth is stressed, and the lack of knowledge on how overloading of the pulmonary circuit is prevented is mentioned. It is necessary to distinguish intraventricular haemorrhages of anoxic origin from subdural haemorrhages of traumatic origin, but, while certain extensive lesions can be recognised, the anatomical changes in lesser degrees of brain damage are unsatisfactorily appreciated. The relationship of perinatal anoxia to the pathological changes found in most cases of brain damage of later childhood also remains uncertain.

RÉSUMÉ

Anatomie pathologique de l'anoxie

Les lésions anoxiques peuvent être consécutives aux vices intrinsèques de l'individu et c'est seulement lorsqu'elles résultent des conditions du milieu et sont extrinsèques qu'on devrait voir en elles, et non dans le vice initial, la cause du décès. Bien que très importantes les causes primaires d'origine maternelle, placentaire et obstétricale de l'anoxie extrinsèque au cours de la période foetale et néonatale ne sont pas discutées ici.

Dans l'anoxie, les lésions principales sont dues à la congestion des voies vasculaires, aux hémorragies des petits vaisseaux sanguins et à l'écoulement du liquide oedémateux. Les lésions varient quelque peu selon les organes. Dans les poumons, on souligne l'importance du contrôle du flux sanguin à la naissance; les auteurs font observer qu'on ignore comment prévenir la surcharge du circuit pulmonaire. Il faut distinguer les hémorragies intraventriculaires d'origine traumatique, mais, tandis que certaines lésions étendues peuvent être diagnostiquées, les modifications anatomiques moins importantes causées à une lésion cérébrale ne peuvent être évaluées de façon satisfaisante. Le doute subsiste aussi sur les rapports entre l'anoxie périnatale et les modifications pathologiques relevées dans la plupart des cas de lésion cérébrale de l'enfance.

ZUSAMMENFASSUNG

Pathologische Anatomie der Anoxie

Anoxische Störungen können durch innerliche Defekte des Individuums bedingt sein und müssten nur wenn sie aus den Umgebungsverhältnissen entstanden und äusserlich sind als Ursache des Todes, anstatt des initialen Defektes, anerkannt werden. Obgleich sehr wichtig, werden hier die primären mütterlichen, plazentalen und obstetrikalen Ursachen der äusserlichen Anoxie während der fötalen und neonatalen Periode nicht erörtert.

Die vorherrschenden Läsionen der Anoxie entstehen durch Kongestion der Blutbahnen, Hämorrhagien der kleinen Blutgefässe und Abfluss der Odemflüssigkeit. Die Läsionen verändern sich etwas in den verschiedenen Organen. In den Lungen wird die Wichtigkeit

der Kontrolle des Blutflusses bei der Geburt betont und der Mangel an Kenntnissen über die Möglichkeiten, der Ueberladung des Lungenkreislaufs vorzubeugen, wird erwähnt. Intraventrikuläre Hämorrhagien traumatischen Ursprungs müssen unterschieden werden, aber, während es möglich ist, gewisse ausgebreitete Schädigungen zu erkennen, kann man anatomische Veränderungen bei geringeren Hirnschädigungen nur ungenügend schätzen. Die Beziehungen zwischen der perinatalen Anoxie und den in den meisten Fällen von Hirnschädigungen der späteren Kindheit angetroffenen pathologischen Veränderungen bleiben auch ungewiss.

REFERENCES

- Drinker, C. K. (1945). Pulmonary Edema and Inflammation. Cambridge, Mass.: Harvard University Press.
Greenfield, J. G., Blackwood, W., McMenemey, W. H., Meyer, A., Norman, R. M. (1958) Neuropathology. London: Arnold.
Grøntoft, O. (1954). 'Intracranial haemorrhage and blood-brain barrier problems in the new-born.' *Acta path. microbiol. Scand.*, Suppl. C.
Morison, J. E. (1952) Foetal and Neonatal Pathology. London: Butterworths.
Obes Polleri, J., Hill, W. S., Matteo, A. L. (1959) 'Ensayo de analisis hidrodinamico de la circulación sanguinea.' *Bol. Inst. puer. Univ. Brasil*, 16, 237.

Attitudes of Children in School

With Special Reference to High Wick, a Psychiatric Unit
for Psychotic Children

MARGARET CHOJKO

Teacher-in-Charge

High Wick is a unit for psychotic children, aged from 3 to 12 years, administered by the Mid-Herts Group Hospital Management Committee. Dr. George Stroh is the consultant psychiatrist in charge. The unit is housed in a pleasant country mansion just outside St. Albans, in Hertfordshire, and is run as a children's home rather than as a hospital. There are at the moment 19 children, all resident. There is a school on the premises to which all the children come for part of each day.

MOST of the children's attitudes which I am about to describe concern those towards school in general, including the teacher, learning and attainment level, and relationships with each other.

A Normal Infant School

Before discussing children's attitudes at High Wick I should like to look at the attitudes of children in a normal school that approximates most closely to the school at High Wick—namely, a class in an infant school. The age-range is much greater at High Wick, including children up to 12 years, but the school is run on infant-school lines and each period is as much like an infant school day as possible. On going into a class in an infant school, the first thing that strikes one is the amount of activity that is going on. The children always seem to be doing things, there is purposeful moving about, noise, chatter and a general air of busyness. The children are playing. It seems that there is a great deal of imitation of grown-up activity. The children are repeating their every-day experiences in the outside world—playing at being mother in the wendy house or shopping in another corner, or simply

being out for a walk around the classroom with their 'children'. They reproduce many of their confusing every-day experiences and by playing them make them understandable and absorb them. This seems one way in which children are able to solve their own problems. I have often watched anxious or worried children play and although I have not always known exactly what was going on, I have seen the same children emerge from a stormy session in the wendy house, relaxed and relieved, having somehow solved their problems by playing them out. The play of young children helps to develop that concentration and absorption which is so necessary to them in learning situations in and out of school.

Another attitude which is essential to a child growing up is his curiosity towards everything around him. Young children are always exploring and investigating; they poke into everything, pull things apart and try to put them together again and generally 'interfere'. The materials presented to them stimulate them to try many things—not only that for which the apparatus was designed. Consequently, there is usually hard wear and tear of

equipment in an infant class and quite a bit of destruction.

The children are very much aware of each other—the very first day in school they will learn each other's names and they very quickly find out lots about each other by questioning and chattering among themselves. They try to gain the approval of the other children—when they do, they gain in confidence and independence.

Young children persevere for a surprisingly long time, particularly at a self-chosen activity, and most children will stick at a puzzle or other piece of apparatus until it is finished.

Most children come to school having the idea that they are going to learn. Parents help in this. Children hear parents saying 'you'll be able to read that when you go to school', or 'how clever you'll be when you go to school', so that most children have accepted the teacher's role and approach their work quite willingly. I found learning problems relatively few, and children who are likely to have difficulties are soon apparent among so many.

The teacher should widen the children's experience and try to bring their make-believe into contact with reality. Her attitude should be that of a 'guide', helping the children to learn by their own experience, rather than being merely a dispenser of information.

The Position at High Wick

I found the attitudes in High Wick very different from those of a group of normal children. My first impression was of some strange sort of isolation. Not that there is no contact at all, but rather that each child seemed to be contained in his own little circle and only if someone from outside broke in was there any contact. There was no spontaneous reaching out to another child or adult, no friendships or group activity. There was none of the bustle or noise of an infant class—for the first time

in my teaching life I wished that the children would chatter, for the quietness was quite strange. The spontaneous imaginative play was missing. One little girl seemed at first to be the exception; she did play in the wendy house with two dolls which she belaboured, but even so she only ever repeated this particular piece of behaviour and did not include any of the other children in her play. Nevertheless it was a reflection of her state of mind. When she went through an unsettled period owing to the arrival of a new baby at home, she stopped playing altogether. Only recently, as she has become more reassured, has she begun to play again, now sometimes including another child as 'daddy'.

* * *

All the children were anxious at the arrival of a new teacher. This was normal enough, but the anxiety was shown in rather abnormal ways—there was a great deal of perseverance and some children took refuge in rituals. The rituals and pre-occupations were completely new to me. I had not met this at all in normal teaching or even with backward children. Some of the rituals were quite obvious. One girl of 9 years had to hold certain objects—always the same ones—which filled her hands and arms, making it impossible for her to use the apparatus in the normal way. She did in fact use her tongue instead, picking up pegs and pictures with the underside and becoming very adept at finishing her tasks in this fashion. Her choice of objects was rather bizarre; it included a small glockenspiel, a hammer, part of a toy telephone, a plastic beaker, two soft covered books and a number of small objects—a toy soldier, bricks and a spoon. I found it impossible to relieve her of any of the objects; if I tried she became extremely agitated, pulling her hair and banging her head. Rather than take away her objects I began to add to them. She

accepted my offerings and each day became more loaded. When I saw she could hold no more I offered her Smarties, the sweets which she loved, and in trying to take them she dropped several of her objects. Once they had been dropped and picked up a few times, she began to abandon them and work freely.

I had a similar experience with another child, this time a boy of 10 years. He had to do the same pieces of apparatus in the same order every day—it had become a ritual for him. If I tried to change this by giving him something before he had finished his ritual, he became very anxious. He would dance on the spot, look away from the thing I was showing him and was unable to tackle it until he had completed the work in his own order. I used the same principle as with the girl: namely, I tried to break the order by adding to it and holding out the promise of something new and enjoyable. So each day I tacked an extra piece of apparatus on to his and rewarded him when he had completed it all. One day he did the things out of order, whereupon I pointed this out to him and the next day he began with the second piece of his apparatus, and after this the whole thing began to break down until he can now start with anything at all without anxiety.

Some pre-occupations are less obvious, however. One boy began to play records every afternoon. I felt quite glad that he had become interested in something at last. However, I noticed that he was very intense about it and that he pushed the other children away where they could not hear the records. Only when the record player was out of action for a few days did I realise that this had become a pre-occupation, for he was unable to do anything else but lie on the floor and worry about how many hours it would be before it was mended. As he was also interested in the records themselves and particularly

My Bonnie lies over the Ocean, I played songs from the records beginning with this one, at singing times. He then became very enthusiastic about singing. His interest is now extending to stories as I have introduced him to 'Hans Andersen' on records. In this way it has been possible to use his pre-occupation to widen his interests, with the result that a few days ago he was able to leave his records for the first time when given a choice between them and singing. I found from these experiences that the teacher must build upon what is there and guide the child's activities towards learning, even if those activities are pre-occupations or rituals.

I had expected to find a great deal of aggressive behaviour, but in fact was surprised to find that I had often received more knocks and bruises during a day in the infant school. There was a certain amount of verbal aggression—but this was only from one or two children. On the whole, aggressive behaviour was conspicuous by its absence. The same applied to destruction of apparatus. The materials did not seem to stimulate many of the children at all, others were obsessively careful with it and so the wear and tear was almost nil. The curiosity which one expects from children was absent. This attitude can be a great strain on the teacher—it seemed at first that many of the children were not aware of me at all and I had the problem of forcing a contact without making the withdrawal even greater.

* * *

None of the children could tolerate frustration. One morning two boys asked me a question at the same time. Instead of answering either of them I spoke to a little girl who had asked me something previously. Within seconds both boys flew into a tantrum, lying on the floor, kicking tables; no answers would pacify them. I have noticed that as a child gets well, it is

in their 'frustration tolerance' that the improvement shows most in school.

Having so many problems and fantasies to cope with makes concentration doubly difficult for the children. Many of them become physically tired after a few minutes' concentration. Others can go on for hours pouring out their fantasies in paint or stories, but when it comes to writing a story about something which they may have done at the week-end, for instance, a few sentences are squeezed out with great anguish and effort.

One or two children, owing to unfortunate teaching experiences or overmuch pressure at home, have a very great resistance to any sort of 'teaching'. These children often suffer from a sense of inadequacy owing to previous failures in the learning situation and this makes them doubly resistant. Often I find that the only way of succeeding with these children is to use another child. For instance, one boy would watch the other children paint but always maintained that he could not do so. When I attempted to help him he really seemed unable to draw even a crude figure. However, a boy whom he admired very much ordered him to draw 'people' on the group painting, to my amazement he succeeded where I had failed—the boy drew several figures and was able to accept a suggestion to 'hold your charcoal like that'. Sometimes I find it necessary to teach by leaning across the child and teaching the one next to him. Often in this way his interest is aroused and he will absorb much of what I am telling his neighbour, especially as he does not feel that it concerns him.

* * *

Not all the attitudes are completely dissimilar from those of normal children. There have been some new entrants recently, some of whom have helped in developing some sort of group feeling.

There are still children who have bizarre patterns of behaviour which they play alone, but there is much more group feeling than formerly among the older children. This is noticeable out of school also: four or five children in a sort of gang interested in what the others are doing and often playing together.

We managed to produce a group painting in which everyone except the four smallest children took some part. I became completely redundant when this was being executed, in marked contrast to a similar effort made some months earlier, where I was needed to give help and persuade the children to paint at all. In the first effort each child painted his own piece and it was difficult to persuade them to allow one child to fill in around another child's piece. This time, however, many of the children saw the picture as a whole and there was quite a lot of enthusiasm and a great deal of interest and pride in the finished picture. This group feeling has shown itself in various ways, none more normal than the following: I left the children working around the table for a few minutes. As I returned I heard some giggling. This still being rather a phenomenon I stopped outside the room to eavesdrop. I heard one voice say, 'I've drawn five Chojko's down the drain' (this from a boy who does not usually draw). A second voice said, 'I've drawn her with her hair wet.' The next little voice said, 'Look, I've put her down a drain with bars on.' There was some more giggling, then as I entered the room there was a scuffle and all the pieces of paper which had been causing so much amusement were pushed under the table. Far from being hurt, I was delighted!

* * *

I feel that many of the attitudes of a teacher in a unit for psychotic children should necessarily be the same as the attitudes of a teacher in a normal infant

school. She must have the same aim of bringing the child's world into contact with reality—to widen his experiences and to help the child to learn. However, the rewards are much slower than in normal teaching and so I think it important that the teacher should be able to take a long-term view of what she might possibly be able to achieve with these particular children. Usually one can forecast roughly what a certain child in an Infant school may have learned in six months' time. This is impossible with the children at High Wick. Their learning often does not conform to any known pattern. A child may progress at a great rate in one particular field—for example in number—for a few weeks or months and then may stop for several months before making any further progress. It seems that many of these children have much longer learning plateaux than normal children, and also periods of regression—i.e. a return to earlier states of intellectual and emotional development.

Unless the teacher realises this she could easily become disheartened. The teacher should be able to see when inferior work is sometimes a sign of progress. For instance, one boy would spend every available minute producing the most complicated paintings, beautiful in colour and composition, yet as he improved his paintings deteriorated until they became the pictures of any seven-year-old painting for pleasure.

A sense of humour, so necessary to any teacher, is essential in High Wick. Many of the children's reactions and tantrums can be very wearing unless one can sometimes see the humour of the situation.

Finally, the teacher should be on the lookout to give help and encouragement in anything at all—especially in the handling of equipment and the learning of skills. Every new thing learnt helps the children towards independence and gives them that feeling of steadiness and confidence which they need so much.

SUMMARY

A comparison is made of school behaviour of normal infants and psychotic children. The most marked difference is the lack of imaginative spontaneous play in the psychotics. The children are often isolated, showing no curiosity towards themselves or their surroundings.

Learning problems are invariably present and varied. Inconsistent concentration and uneven development show in some children by their ability to acquire skills far ahead of the rest of their attainments. Regression from already acquired behaviour patterns, often seen for short periods in normal children, is often severe in the psychotic; and learning plateaux—a normal feature of learning—are much more pronounced in children at High Wick.

There is a great deal of anxiety shown in extremes of behaviour not usually observed in normal children, such as perseverance, obsessional and ritualistic behaviour. On the whole aggressive behaviour is conspicuous by its absence. Frustration tolerance is extremely low and it is here that any improvement first shows itself.

Once a relationship is established between the teacher and the children, similarities begin to be apparent. One can stimulate group activities, and reactions to group situations and other children become more normal.

RÉSUMÉ

Attitudes de l'enfant à l'école

On a comparé le comportement scolaire d'enfants normaux et d'enfants psychotiques. La différence la plus marquée est l'absence du jeu imaginaire spontané chez les psychotiques.

Les enfants sont souvent isolés, ne montrant aucune curiosité vis-à-vis d'eux-mêmes ou de ce qui les entoure.

Les problèmes d'acquisition sont invariablement présents et divers. L'inconstance dans la concentration et un développement inégal se manifestent chez certains enfants par leur facilité à acquérir certaines spécialisations, bien au-delà du reste de leurs acquisitions. La régression de certains aspects déjà acquis de comportement, que l'on constate fréquemment pendant de brèves périodes chez les enfants normaux, est souvent grave chez les psychotiques; et les paliers dans leur instruction, qui sont un trait normal de toute instruction, sont plus prononcés chez les enfants de High Wick.

On observe un degré prononcé d'inquiétude, manifestée par des extrêmes du comportement tels qu'on n'en observe généralement pas chez les enfants normaux, par exemple persévérance, comportement obsessionnel et rituel. Dans l'ensemble, l'agressivité du comportement est remarquablement absente. La tolérance à l'échec est extrêmement faible et c'est sur ce point que se manifeste d'abord une amélioration quelconque. Une fois établis les rapports maître/enfants, des ressemblances commencent à apparaître. On peut stimuler des activités de groupe, et les réactions vis-à-vis des situations collectives et des autres enfants deviennent plus normales.

ZUSAMMENFASSUNG

Verhalten des Kindes in der Schule

Ein Vergleich zwischen dem Benehmen normaler und psychotischer Kinder in der Schule wird gemacht. Der hauptsächlichste Unterschied ist der Mangel an freiwilligem Einbildungsspiel bei den Psychotikern. Die Kinder sind oft einsam und zeigen keine Neugier gegenüber sich selber oder ihrer Umgebung.

Lernprobleme sind beständig vorhanden und verschieden. Unbeständige Konzentrationsfähigkeit und ungleiche Entwicklung offenbaren sich bei manchen Kindern durch ihre Fähigkeit, Geschicklichkeit in einigen Fächern zu erreichen, weit über ihre übrigen Erwerbungen hinaus. Regression der schon erworbenen Verhaltensarten, die oft kurze Perioden lang bei normalen Kindern beobachtet wird, ist oft bedeutend bei den Psychotikern; und die Lernstufen—eine normale Eigenheit des Lernens—sind viel ausgeprägter bei den Kindern aus High Wick.

Man bemerkt starke Ängstlichkeit, die sich in Extremen des Verhaltens, die man gewöhnlich nicht bei normalen Kindern beobachtet, zeigt, zum Beispiel Perseveranz, Zwangs- und rituelles Benehmen. Im ganzen ist aggressives Benehmen auffallend durch seine Abwesenheit. Frustrationstoleranz ist sehr schwach und in diesem Gebiet macht sich irgendeine Verbesserung zuerst bemerkbar.

Sobald eine Beziehung zwischen dem Lehrer und den Kindern entstanden ist, werden Ähnlichkeiten sichtbar. Es ist möglich, den Gruppentätigkeiten einen Antrieb zu geben und die Reaktionen auf Gruppensituationen und andere Kinder werden normaler.

Provision of Special Facilities for 'Ineducable Spastics'

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AT Harperbury Hospital we treat very severe cases of cerebral palsy with mental defect, who have little chance of becoming independent even in the specialised environment of a mental deficiency hospital. We are not very fond of the term 'ineducable', because we believe that much can be achieved in these cases and that there are very few who cannot benefit materially from skilled care and treatment. To encourage both parents and staff we prefer to describe our cases as suffering from severe multiple (or special) handicaps, requiring special care and attention.

For a long time we in mental deficiency have lagged behind those working with 'spastics' of normal intelligence; but in fact there was little that could be done for our type of case until a few years ago, since all available facilities and trained personnel were fully occupied. The usual Cerebral Palsy Units would not look at our patients, expecting normal or superior intelligence in their candidates for treatment. This also applied to severe neurological disabilities. I have never been successful in my attempts to find places in a 'Spastics' Unit for the severely affected athetotics of almost normal intelligence whose neurological disabilities severely affected their speech.

At Harperbury, Dr. Taylor and I have felt that—because of its importance and the implication of different disciplines—the problem could only be tackled effectively as a major project. This led to our proposal that an experimental unit should

be set up for the treatment of our patients and the investigation of their disabilities. We have finally achieved this end, thanks to the Nuffield Trust and the North-West Metropolitan Regional Hospital Board.

The Experimental Unit*

The new Unit is a building with two large rooms—one for physiotherapy, and the other for occupational therapy, education, and cine photography for clinical records. There will be enough office accommodation for the consultant to examine cases, with a room for psychological testing and a room for speech therapy, with space for, we hope, audiometry. We shall have to forgo EEG facilities for the time being, because of lack of money.

The staff, when we get our full complement, will consist of a consultant physician—rather more than half time—a psychologist, 3-4 physiotherapists, a speech therapist, a teacher, and one or two occupational therapists. The psychologist, the occupational therapists and especially the teacher will work in close contact with existing departments. The Unit will be for day treatment; the patients will come from the wards in the morning and return there after spending half to a full day in the Unit.

The type of case we have to deal with

* Since this paper was read, the Unit has been in operation for a year, under Dr. Karel Bobath's direction. We hope to describe the work of the Unit and the results obtained in a later paper. A.S.

forms, of course, a continuum with the 'normal' type of spastic, and in so far as it differs it does so by an associated and variable mental defect, which usually can be ascribed to damage to the central nervous system; it must be remembered, however, that there is often a lack of correlation between intelligence and severity of paresis. In many cases speech is severely affected and both neurological and mental factors combine to produce this defect. A closely associated problem which we find in low-grade patients—usually idiots—is that of disturbed mobility without any signs of neurological damage. Grace Woods (1957) refers to this type of case under the heading of 'rigidity' and assumes that there is neurological dysfunction; I personally feel that the condition may be due to psychological factors.

Since these cases have a generally similar disability and sometimes require similar treatment, it is convenient to have a blanket term for them, but one should not forget that both aetiologically and clinically they are individual and distinct conditions, and because they have a common name they are no more identical than are the conditions that go to make up mental deficiency.

Are we justified in expending all this effort in trying to help very severe cases with the additional disabilities of mental defect and speech disorder? I believe that we are.

First, the bedridden and contracted neglected cases of cerebral palsy are most cruelly handicapped and present a severe nursing problem; anything that can be done to alleviate their deformity and help with their nursing is amply worth while.

Secondly, the emotional benefits which follow the mobilisation of a patient, making him independent of other people for his immediate and intimate needs, such as feeding, dressing, going to the lavatory, etc., can hardly be exaggerated.

I should like to illustrate this point by quoting the case of an adolescent with spina bifida and myelocoele, and a medium amount of mental defect. He suffered from trophic ulceration of his denervated lower limbs and chronic osteomyelitis, which kept him confined to bed and necessitated recurrent operations. His behaviour throughout this period was very difficult: he was asocial, troublesome, emotionally labile, and subject to outbursts of disorganised behaviour, when he screamed, smeared faeces, and was quite intractable. It was finally decided to amputate his legs and he was fitted with prostheses. He has been walking on his artificial legs for some months past, and the change is almost unbelievable. He functions many years above his previous social level, his relationship with people has improved, and he is now friendly and co-operative. Above all, he has generally shown much greater interest in his surroundings, working well and happily in occupational therapy, and has now expressed a wish to learn to read. His intelligence, as measured by tests has also gone up.

Thirdly, we hope that treatment will enable our patients to cross the dividing line between being a chronic hospital patient and one with sufficient independence to return to life in the community—in some cases the difference is not so great as would at first appear.

The final, and in our opinion most important, reason is that we believe we should explore the relationship between early physical handicap and mental development.

Effect of Physical Condition on Mental Development

Schilder and Loretta Bender, among others, have stressed that the mental development of a young child is a function of learning by exploration and experience of its environment in relation to its ego and its body. The child with cerebral palsy suffers from a double handicap, being not only restricted in mobility but also having special sensory disabilities and other diffi-

culties in co-ordinating sensation with motor activity. The classic case is the child with persistent tonic neck reflexes, who has to turn his head away from an object which he is trying to grasp, thus being able to learn only with difficulty to co-ordinate visual and sensory sensations.

It is to test this thesis that we feel justified in setting up an experimental Unit; for, if it is successful, early and intensive treatment may save patients from having to spend their lives in hospitals for the mentally defective.

A corollary to this enquiry would be the investigation of the presence or absence of *critical maturation periods* during the development of patients. It would be important to establish the prognosis of adults who receive skilled treatment. This is a particularly important question at present in the field of mental deficiency, where there are a large number of adults in need of treatment who have been neglected in the past. It would be essential to establish how much mental improvement could be expected *pari passu* with the alleviation of their physical condition. These critical periods are of course very important in the development of young animals, as has been amply demonstrated by the ethologists.

The setting up of an experiment to assess the variation in mental development produced by treatment will require careful thought and preparation. The problem will be largely psychological and will necessitate the elaboration of tests with a predictive value for the results of treatment. These will differ from ordinary tests of intelligence, which test the level of mental attainment; we are concerned with picking out those pointers which will indicate that the level that has been attained is not permanent but is capable of being altered by treatment. Such tests can only be developed during the course of treatment, operationally, and as I see it we

shall have to start with adaptations of existing tests.

The question of *intelligence testing* of spastics is a difficult one, and all psychologists of experience tend to adapt the usual tests by flexibility in their administration. In ordinary intelligence testing, only the correct answer is acceptable, and the tester is not able to credit the answer, if not formulated correctly. In testing spastics, however, it is essential that the tester should be able to guess the response. The *Binet* test is still liked by very experienced psychologists: Sarason, Dunsdon and Eleanor Schonell have described, for example, modification in administration and selection of subtests which are most useful in the testing of spastic subjects. I have no personal experience of the *Columbia Mental Maturity Test* but there is a sharp division of published opinion on its usefulness. The criticism that it does not allow for perceptual disabilities of the children is one which must be applied with even greater cogency to a lot of test material used, such as the *Progressive Matrices*, where discrimination of shape and figures is particularly important. *Ammon's Full Range Picture Test* does not in my short experience of the test recommend itself, as the pictures are not clearly drawn, and many responses refer to the same picture, thus encouraging perseveration.

My psychological colleagues at Harpersbury have been trying out the *Peabody Test* which consists in picking out of four pictures the more appropriate to the stimulus word. In tests with non-spastic imbeciles it gave a very high correlation (0.74) with the *Binet* (Terman-Merrill) and was easy and quick to administer. We hope to try it out on our cases of cerebral palsy in the near future. The test, incidentally, is still being standardised.

It may be that an assessment of personality and of behaviour under test

conditions will be the most useful prognostic indication of the effects of treatment, but formal testing will allow the elucidation and measurement of specific disabilities.

Assessment of Results of Mental Testing

Two special factors must be kept in mind when assessing the results of mental tests in this type of patient: (1) the possible presence of special sensory disabilities, of a higher integrative order, such as the different agnosias, spatial disorientation, and disturbances of the body image, which will all require special elucidation; and (2) the presence of speech disturbance. The question of *speech disturbance* in spastics with mental disability is of great practical and theoretical interest. Luria and Yudovich (1959), developing Vigotski's earlier work, emphasise that the acquisition of language is an important factor in the development of the capacity for abstract thinking and symbol formation. In the more intelligent cerebral palsied patient, neurological defect does not seem to affect the acquisition of internalised speech and comprehension, even in cases which are completely dysarthric; in the patient with a double handicap, however, the inability to verbalise may affect the discrimination of words and retard the development of internal speech. The relation of speech to development and to intelligence is a very involved and controversial problem.

Returning to the *special disabilities*: the diagnosis of special sensory and apraxic deficits is most important when dealing with cases with mental retardation, since their effects may be mistaken for mental defect, and in mentally defective patients the importance of each factor must be estimated.

Defects such as blindness and obvious deafness are easily diagnosed. Partial deficits, such as high-tone deafness, are much more difficult, yet by interfering with the comprehension of speech this may

mimic intellectual defect. According to Grace Woods (1957), the athetotics seem particularly prone to this. It is well known that deafness and athetosis are found together, and Richards has described cases where both conditions were genetically determined. Ideally all our cases should be screened by audiometry but this if of course a counsel of perfection. Mary Sheridan (1958), however, has devised a series of hearing tests applicable even to very young children and low-grade cases. These consist of a series of familiar noises graded for pitch—the rattle of a spoon in a cup, the crumpling of paper, the ringing of bells, and so on.

When we come to the higher integrative functions, such as *agnosias*, *aphasias* and *apraxias*, I have an uneasy feeling that we physicians have tended to ignore them and it was the educators who stressed their importance. In the course of teaching, agnostic and apraxic symptoms should obviously be much in evidence as learning difficulties. Frequent complaints are the inability to recognise and distinguish shapes (including letters) and directions and to translate two into three dimensions—i.e., to interpret pictures.

The *concept of number* is particularly difficult for these children, and this is frequently associated with the other symptoms of the Gerstmann syndrome, with finger agnosia and loss of laterality.

Disturbances of the body image are common and manifest themselves as imperception of affected parts of the body. However, some of the tests of body image imperception are also tests of visual spatial orientation and inability to draw human figures or put together object assembly tests may not necessarily be due to the disturbance of the body image.

On the motor side, apraxia plays its part in causing clumsiness and other motor disabilities, as well as paresis. And in some cases inability to speak is apparently due

to apraxia of the motor apparatus of the mouth, throat and larynx. It is impossible to discuss these disabilities beyond drawing attention to their importance and to the need to diagnose them.

Personally, I feel strongly that though aphasic symptoms are most evident in an educational psychological setting, dealing with them should be considered largely a neurological problem. The presence of epileptic and convulsive disorders often determine the symptomatology of these defects, and can also markedly affect personality structure and behaviour patterns. Many of the disabilities of cerebral palsied patients were ascribed by the Gestalt school—Goldstein (1948) and his followers in the educational field, Straus *et al.* (1947), Warner and others—to loss of cerebral tissue. This loss not only determines the deficit in cerebral functioning but deprives the total individual organism of the ability to deal with environmental threatening situations. An imperfect adjustment follows which produces the great rigidity in habits and behaviour; and when these are unable to protect the individual from demands made by his environment he erupts into uncontrolled irritable tantrums which Goldschtein has described as 'catastrophic reaction'. Loretta Bender devoted her interests to the study of the personality of children with brain injury, particularly when it occurred later in life, such as the encephalides following virus infections, severe burns, and other acute encephalopathies. She too related the dependent egocentric clinging and irritable personalities which these children presented to imperfect attempts at adjustment.

The importance of this theoretical point of view is that it determines the programme of treatment. The patient is trained to make the most of his remaining abilities to circumvent his difficulties; essentially the same methods are used as those in training traumatic aphasia.

However, the successful results of neurosurgery, particularly of hemispherectomies, show that the symptoms of behaviour disorder may be due not just to a negative symptomatology due to cortical deficit, but may be a positive symptom caused by the disruption of normal functioning by the abnormal neuronal discharge. Of the patients who have been subjected to hemispherectomy, most have shown marked disturbance of figure background, discrimination, perseveration, hyperexcitability and explosive emotional reactions to minor stimuli. All these symptoms vanish after successful hemispherectomy, and, particularly in the more intelligent patients, learning and education become possible where previously they had been severely disrupted by the symptoms referred to. The series of cases operated on by Cairns and analysed by Miss Davidsen (1951) are a good illustration. Some became educable, one finished school, and one went to university. It is well known now that successful hemispherectomy checks both epileptic seizures and also dysrhythmias originating in the affected hemisphere which has been removed.

Although the effects are well known, I personally have not seen any attempts to alter the existing theories to fit these facts. The practical consequences are of course important in that they affect the rationale of treatment.

Further, if, as the Gestalt school claim, the symptomatology is due to deficit, training is the answer; if, however, it is due to abnormal cerebral functioning originating in pathological foci, then the development of neurosurgical techniques for dealing with these lesions is of urgent interest. I am of course discussing not only the hemiplegias but all types of cerebral palsy showing these symptoms. I have the strong impression that many of the features of the personality structure and behaviour patterns in our cases are

due to subthreshold convulsive phenomena. Any personal assessment should be done in close association with the evaluation of the state of the central nervous system and the motor and sensory factors, which may play an important part in forming the total personality picture. Unfortunately, the facilities for EEG studies in mental deficiency hospitals are now conspicuous by their absence, but in time they will certainly be considered as essential for the examination of defective 'spastics' as radiological investigation is now.

Psychodynamic theories are of the highest importance in determining personality structure, but there should be no incompatibility between the organic and psychopathological approach; the two methods should be complementary. Indeed, as Schilder (1931), in *Brain and Personality*, showed many years ago, investigations of the boundaries between mental and physical phenomena are not only of extreme interest but are concerned with fundamental questions of cerebral and mental functioning.

Psychological Factors

The psychological aspect is the most important single factor, for it determines whether the patient can come to terms with his disability and affects his ability to make a satisfactory adjustment.

In general, the patient's attitude to his disability will depend on his personality and the need to adjust to the handicapped existence by a number of reactions. Most children will show an accentuation of normal dependence and insecurity, which will be accompanied by emotional immaturity. In the community at large they will find themselves excluded from the group of normal children, if only because they are physically incapable of keeping up with the others. The child will be thrown much more intensely on the emotional relationships within the family; and the attitude of

the family will have a greatly magnified effect on the child and his personality.

The situation is much modified if the child is brought up in a hospital atmosphere. The well-known effects of hospitalisation on children are somewhat modified by the greater acceptance of physical and mental disabilities. The emotional relationships which the child can form are largely a matter of circumstance and luck.

The junior nurses tend to succeed each other fairly rapidly in the course of their training, but the sisters and staff nurses can provide stable and dependable relationships, even though they never achieve the warmth of feeling with the majority of children that at their best would be found in a family group. The situation is, however, alleviated by the 'adoption' of children by adult patients working in the wards, and by some nurses. This can be a great encouragement, and often determines the rate of a child's physical progress. Sometimes a young child starts walking once he has established a sufficiently good relationship with one of the adults who is prepared to devote time and attention to him. It is essential for all children to have sufficiently warm and secure relationships in hospital.

The effect of mental defect on 'spastic' children is variable, and can produce either a general emotional flattening and indifference to surroundings or emotional instability and irritability. The emotional needs and problems of the mentally defective child with cerebral palsy in an institution are just as important and pressing as they are in a cerebral palsied child of normal intelligence. Where indifference is the presenting clinical picture, there is a great need to combat the withdrawal of the patient from his surroundings, and to stimulate interest in group activities. There is danger of these patients being allowed to sit about

without any occupation and to suffer in consequence.

In the severely affected child careful assessment of intelligence is of great importance, for many who are very severely handicapped and whose speech is completely incomprehensible are in real danger of being classified as low-grade imbeciles or idiots, and not given the benefit of treatment. It is striking how some children capable of very limited activity will concentrate on it with great intensity—for example, one severely handicapped child with just the use of the fingers and wrist of one hand spends his time spinning tops and toys on the floor, and has developed a high degree of skill at this occupation.

The correlation of personality structure with a specific physical lesion has not received the attention it deserves. The athetotics, for example, show marked emotional lability which may be of hypothalamic origin, and at the same time they possess charming personality and are capable of establishing excellent personal relationships. On the other hand, some epileptics develop the typical personality picture associated with this condition. Its existence has been strongly denied, but it is much more likely to manifest itself in the setting of a mental deficiency hospital than in patients whose intelligence has been sufficiently well preserved for them to be capable of independent existence in the community.

Methods of Treatment

Physiotherapy

I am convinced that the fundamental form of treatment in cerebral palsy is physiotherapy. I had been used to seeing these cases become increasingly contracted and crippled in spite of muscle relaxants, passive movements, extension, and repeated operations for the unavailing lengthening of tendons. It was therefore a revelation to

find how much can be achieved by modern physiotherapeutic methods, when I had a chance of studying the Bobaths' film records and watching Mrs. Bobath's demonstration on my own cases; cases which I was convinced had permanent contractures were shown to be suffering from spasm which could be abolished on the couch in a few minutes.

There are of course many methods of physiotherapy based on different underlying principles. Phelps (1958) has summarised them as follows:

1. Conditioning according to Pavlov's principles to establish reciprocal motion, bearing in mind Sherrington's law of reciprocal intervention (used by Phelps).
2. Relaxation and motion from the relaxed position following the principles of Jacobson and other relaxation methods (used by Phelps).
3. Increasing awareness of contraction or joint and muscle Kinaesthesia by resistive therapy (Kabat).
4. Utilisation of residual patterns both voluntary and semi-automatic, and utilisation of pathological reflexes (Fay).
5. Inhibition of abnormal reflex patterns and facilitation of normal automatic reaction in their developmental sequence (Bobath).
6. Stimulation of contraction and consequent relaxation of antagonists (Rood).
7. Methods of Pohl, Swartz, Deaver and Collis.

One important aspect must be remembered in trying to assess whether a particular method is suitable for the treatment of the mentally defective spastic patient; this is whether the method depends on the patient's co-operation, or whether it can be applied to an unco-operative and, indeed, even to a resistant

patient. From that point of view my colleagues and I have been highly impressed by the Bobaths' method.

Hemispherectomy

I have already mentioned the psychological results of hemispherectomy and its relation to the underlying convulsive phenomena. Marked personality changes can also follow a successful operation. The effect is remarkably similar to that of leucotomy. Patients who had been impatient, irritable, and so impulsively violent as to be considered a danger to other patients and staff have become friendly, tolerant and relaxed. Sometimes this process has gone beyond a satisfactory result and has produced apathy and listlessness, so that the patient is difficult to interest in any activity and if left to himself sits about completely inactive. This again has been described as a sequel of leucotomy.

Altogether, 8 hemispherectomies have been done on Harperbury patients; the operations were done at Queen Square and reported in their series and also by Fleischhacker (1954). There was no immediate postoperative mortality, but two deaths should be ascribed to the operation. A child with the Sturge-Weber syndrome died a couple of months after the operation, and another patient died after surviving for a year, during which she became increasingly listless and retarded, the running down process affecting not only her mental but also her physiological functions. Comparing our series with the published cases, I have the impression that the improvement is definitely correlated with intelligence and the presence of symptoms indicating disturbed function, whether manifesting itself in sensory and psychological disabilities, or in personality and behaviour disorders. The low-grade case has a poorer prognosis, largely because he is much more likely to have

lesions on the remaining side, masked by the symptoms produced by the affected side, which are discovered at operation. The fits may recur and there may be a deterioration in the personality improvement. In assessing this type of case for operation, one should be guided by the severity of the fits and other disabilities, and above all by how distressed and unhappy the patients are because of their irritability and low frustration tolerance. Some of our higher grade patients have commented spontaneously on how much more contented they felt after the operation.

On the whole, when the fits have recurred and there has been a deterioration in behaviour, the patient's condition has always been better than before the operation.

Operations to remove smaller epileptogenic foci do not appear to have been done in defectives with associated neurological disabilities, and the cerebral injury in our type of case is presumably too widespread to be susceptible to focal operations. Temporal lobectomies for limbic lobe epilepsy have not been done to my knowledge on defective 'spastics'.

Chemothalamotomy

It was after the last war that the modern techniques of inflicting selective lesions on the basal ganglia were developed, particularly by the perfection of stereotactic instruments which allowed electrocoagulation or chemical destruction of selective areas of the basal ganglia.

At Harperbury we have so far had one case only subjected to a chemothalamotomy—but without worthwhile results. I feel, however, that the operation should be given a full trial—if we can find a neurosurgeon with sufficient time to do so! As success has so much to offer, I consider that we are justified in subjecting patients to an operation even if success cannot be guaranteed, especially as with modern

techniques the operation appears to be quite safe.

Even in cases treated with neurosurgery, physiotherapy has an important role to play in accelerating recovery of function.

General Management of Cases in Hospital

It is important that patients should be subjected to a therapeutic atmosphere the whole time, and we plan to integrate our special unit into the hospital life. We hope to keep all our spastics together in special wards and to have not only the patients but also the physiotherapists, the occupational therapists and the nurses moving freely between Special Unit and Ward.

The important role of the nurse in the patient's life is often taken for granted when planning special treatment or special units, and we feel it essential that she should be an integral part

of the therapeutic team. We hope to give instructions to nurses in the Unit and to leave them with the responsibility of continuing in the ward the treatment and training begun in the Unit.

We have a school for our children at the hospital and the instruction in the Unit and the school will also have to be closely co-ordinated.

I think the Unit will be useful to the hospital in more ways than training and treatment of neurological disabilities. Audiometric investigation will be useful in investigating cases without neurological disabilities, and a speech therapist will be of value to many types of patient.

When the Unit is fully developed we shall be prepared to take cases from other hospitals as outpatients, and to put our facilities at their disposal.

SUMMARY

This paper indicates the lack of facilities for cerebral palsy patients in the mental deficiency hospitals and describes plans for the setting up of a Unit to remedy these deficiencies. The building has been made possible by a grant from the Nuffield Trust. The Experimental Unit will be set up as an integral part of Harperbury Hospital. It will consist of two principal rooms, one for physiotherapy and one for occupational therapy and educational activities. There will also be a sound-proof room for speech therapy, and small rooms for psychological testing, examination, etc.

The Unit will be staffed by a part-time consultant, 3-4 physiotherapists, a speech therapist, a psychologist, a teacher and one or two occupational therapists. It will be run as a day Unit, drawing patients from all the wards in the parent Hospital, though it is hoped to concentrate those being treated in special wards.

In contrast to the ordinary type of Cerebral Palsy Unit, Harperbury will have to deal with a large number of adults. The problems of treating defective cerebral palsy patients will need special study, such as the devising of particular predictive tests suitable for their intelligence range and the development of prognostic criteria, the study of the relationship of physical handicap to mental handicap, and the effect of speech development on mental and physical states. The sensory disabilities of these patients will make it necessary to apply special treatment and educational methods.

Finally, surgical methods will have to be integrated with medical treatment.

RÉSUMÉ

Création d'un équipement spécial à l'intention des spastiques inéducables

L'auteur fait état, à la fois, du manque de dispositif rencontré dans les hôpitaux pour oligophrenes pour le traitement des infirmes moteurs cérébraux et des projets concernant la création d'un centre qui remédie à ces lacunes. La construction n'en fut rendue possible

que grace a une subvention du Nuffield Trust. Ce centre pilote fera partie intégrante de l'Hôpital Harperbury. Il sera constitué d'un bâtiment sans étage de deux pièces principales, l'une consacrée à la physiothérapie, l'autre à l'ergothérapie et aux activités éducatives, avec une pièce spéciale insonorisée pour la thérapie de la parole. Le personnel comprendra un consultant à temps partiel, 3 à 4 physiothérapeutes, un thérapeute du langage, un psychologue, un enseignant et un ou deux ergothérapeutes. Le Centre fonctionnera de jour et les malades viendront de tous les services de l'hôpital bien qu'on espère grouper par la suite tous les malades en traitement dans des salles spéciales.

A l'encontre des centres habituels, le Centre de Harperbury prendra en charge de nombreux adultes.

ZUSAMMENFASSUNG

Gründung einer speziellen Einrichtung für unerziehbare Spastiker

In diesem Artikel erwähnt der Autor den Mangel an geeigneten Einrichtungen für die Behandlung der Patienten mit Zerebrallähmung in den 'M.D. Hospitals' und beschreibt die Pläne für die Gründung einer Einheit, um diesem Mangel abzuhelpfen. Diese Versuchseinheit wird im Harperbury Hospital eingerichtet und dem Hospital vollständig eingegliedert sein. Sie wird aus einem einstöckigen Gebäude mit zwei Haupträumen, von denen einer für Physiotherapie, der andere für Ergotherapie und erzieherische Tätigkeiten benutzt werden. Bestehen die Einheit wird mit einem speziellen schalldichten Raum für Sprachtherapie versehen sein.

Das Personal wird sich aus einem Konsultanten, 3 bis 4 Physiotherapeuten, einem Sprachtherapeuten, einem Lehrer und einem oder zwei Ergotherapeuten zusammensetzen. Die Einheit wird bei Tage funktionieren und die Kranken werden aus allen Teilen des Hospitals kommen, obgleich man hofft, später alle Kranken, die sich in Behandlung befinden, in speziellen Abteilungen zu konzentrieren.

Im Gegensatz zu den üblichen Einheiten, wird sich die Einheit von Harperbury mit zahlreichen Erwachsenen zu befassen haben.

REFERENCES

- Ammons, R. B., Ammons, H. S. (1948) The Full Range Picture Vocabulary Test. New Orleans: Ammons.
- Bender, L. (1938) A Visual Motor Gestalt Test and its Clinical Use. American Orthopsychiatric Association. Research Monographs, No. 3. New York.
- Bobath, K. (1956) Persons. communication.
- Burgemeister, B., Blum, L. H., Lorge, I. (1953) Columbia Mental Maturity Scale. New York: World Book Co.
- Cairns, H., Davidson, M. A. (1951) 'Hemispherectomy in the treatment of infantile paraplegia.' *Lancet*, ii, 411.
- Cooper, I. S. (1959) 'Chemopallidectomy and chemothalamectomy for Parkinsonism and dystonia.' *Proc. roy. Soc. Med.*, 52, 47.
- Dunn, L. M. (1959) The Peabody Picture Vocabulary Test. Nashville: G. Peabody College for Teachers.
- Dunsdon, M. I. (1952) The Educability of Cerebral Palsied Children. London: Newnes.
- Fleischacker, H. H. (1954) 'Hemispherectomy.' *J. ment. Sci.*, 100, 66.
- Goldstein, K. (1948) Language and Language Disturbances. New York: Grune and Stratton.
- Horsley, V. (1909) 'The function of the so-called motor area of the brain.' *Brit. med. J.*, ii, 125.
- Luria, A. R., Yudovich, F. (1959) Speech and the Development of Mental Processes in the Child. London: Staples Press.
- Phelps, W. M. (1938) In Illingworth, R. S. Recent Advances in Cerebral Palsy, p. 251. London: J. and A. Churchill.
- Richards, B. W. (1950) 'Congenital double athetosis, deaf mutism and mental deficiency.' *J. ment. Sci.* 96, 280.
- Schilder, P. (1931) Brain and Personality. New York.
- Schonell, F. E. (1956) Educating Spastic Children. Edinburgh: Oliver and Boyd.
- Sheridan, M. D. (1958) 'Simple clinical hearing-tests for very young or mentally retarded children.' *Brit. med. J.*, ii, 999.
- Spiegel, E. A., Wycis, H. T., Baird, H. W. (1958) 'Long-range effects of electropallidoanotomy in extra-pyramidal and convulsive disorders.' *Neurology*, 8, 734.
- Straus, A. A., Lehtinen, L. E. (1947) Psychopathology and Education of the Brain-Injured Child. New York: Grune and Stratton.
- Woods, G. E. (1957) Cerebral Palsy in Childhood. Bristol: Wright.

The Management of Seizures in Infancy and Early Childhood

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CONVULSIVE seizures are a commonplace for those who have the medical care of children. This apparent predisposition of the young to convulsions, which has been recognised since the days of Hippocrates, has resulted in a familiarity which has bred if not contempt at least a rather unwary relegation of infantile convulsions to the group of natural discomforts of childhood such as teething and the exanthemata. This has been largely reinforced by the theory of their production in childhood which regards the immature nervous system as responding by a convulsion to a physiological or pathological stimulus which in a mature nervous system would be ineffective. Thus the concept of the 'teething fit' and the febrile convulsion both imply that under certain circumstances convulsions are to be expected in children and therefore are no cause for concern. The corollary that they therefore require neither investigation nor treatment has not infrequently been drawn, and the comforting assumption that such seizures are not 'epileptic' has tended to withhold this group from neurological attention until the persistence of seizures in later life has demonstrated that some of them in fact are epileptic.

The relationship of infantile convulsions to epilepsy has in the past been the subject of much and often vehement controversy, as often happens when argument belongs to semantics rather than medicine. The relevance of this to the management of seizures lies, of course, in the question

whether casual seizures in infancy, for example, or convulsions at the onset of an infection in early childhood, require any treatment or investigation at all.

The Jacksonian concept of paroxysmal neuronal discharge as the mechanism of the seizure, and its site of origin and direction of spread as determining the manner of its onset and evolution, is the basis on which our present day views are founded. If we regard the basic tendency to paroxysmal discharges as a genetically determined seizure threshold, it follows that factors which lower the threshold will tend to produce seizures while factors which raise it will tend to stop them. There are many factors which lower the threshold and stimulate paroxysmal neuronal discharge, thereby provoking seizures, and these range from cerebral inflammation, haemorrhage or trauma to metabolic abnormalities such as hypocalcaemia and pyridoxine deficiency. Factors which reduce the tendency to paroxysmal discharge and raise the threshold include the various anticonvulsant drugs and also the gradual maturation of the central nervous system from birth to early childhood.

While, therefore, it is probably true that the bulk of convulsions in late infancy and early childhood are the result of the stresses of childhood acting on a temporarily immature nervous system, and that the prognosis regarding recurrences is reasonably good, this conclusion should not too readily be accepted without investigation.

Rational management of convulsive seizures in childhood wholly depends on accurate diagnosis; the aim therefore is to establish as precisely as possible the pathogenesis of any seizure encountered. It is clearly beyond the scope of a paper such as this to consider the multitude of causes which may result in a seizure. Suffice to say that adequate treatment depends on precise diagnosis, which must include the recognition of any underlying cause. Nevertheless, even a detailed investigation may throw no light on the aetiology of the seizure and treatment will therefore remain empirical.

Management of a Major Seizure

There is something atavistic in the horror with which natural parental anxiety is imbued when a major seizure occurs in a child, and the consideration of its cause should undoubtedly be postponed until the immediate treatment of the fit has been applied. It should be mentioned that the evidence that a hot bath has any anti-convulsant effect is negligible—such evidence as there is (Wegman 1939) points the other way, yet tradition dies hard and many a febrile convulsing infant is scalded in deference to it. It seems unlikely that this treatment can do anything but harm.

Immediate sedation is the most effective treatment, and for this several drugs have been advised. For fits in the newborn my practice is to use chloral hydrate, which in adequate dosage is safe and effective. This drug is given by mouth, through a tube if necessary, starting with a single dose of $\frac{1}{2}$ grain per lb. (15 mg. per kg.) body-weight followed by gr. 1 to $1\frac{1}{2}$ (60–90 mg.) four-hourly. Undue sleepiness is an indication for reduction in dosage and there are no other serious side-effects. It is important to reduce the dosage gradually over the next week or two rather than stop sedation abruptly. In older infants one may start with chloral hydrate gr. $\frac{1}{4}$ to $\frac{1}{2}$ per lb.

(8 to 10 mg. per kg.) body-weight as a single dose and follow with gr. 1 to 3 (60 to 200 mg.) four-hourly. Beyond the first year of life I favour, as a simple safe sedative, intramuscular paraldehyde. This is available in 5 ml. ampoules and can be given to children over a year old in doses of 1 ml. per stone (14 lb., 7 kg.) body-weight. This hybrid scheme of dosage is justified on grounds of simplicity, for it is fairly easy to guess the weight of a child to within half a stone, especially if it is remembered that the average two-year-old weighs about two stone and gains thereafter roughly $\frac{1}{4}$ of a stone per year.

Febrile and other Potentially Epileptic Convulsions

Intermediate between seizures which are the direct result of cerebral stimulation by a known pathological process and those which occur spontaneously are those usually termed febrile. It is doubtful whether febrile convulsions warrant separation as a separate category: where a child has a convulsion at the onset of a febrile illness and specific causes have been excluded, the seizure can indeed be called febrile but its pathogenesis is probably a matter of seizure threshold: indeed it is a useful practical point in determining the prognosis of a convulsion to assess the degree of provocation. Where, for example, the convulsion occurs at the onset of an acute illness such as measles it does not necessarily indicate a low seizure threshold and there is therefore no reason to expect a recurrence. When the convulsion accompanies a mild upper respiratory infection with only the slightest pyrexia it should be regarded as being closer to a spontaneous seizure with the implication of a low seizure threshold and liability to recurrence. Repeated convulsions, however, even when undoubtedly 'febrile', are better treated like any other repeated seizure and regarded as potentially epileptic. Indeed,

when the rarities are excluded, what remains is the bulk of cases and in these no causal factor can be found: to these it is logical to apply the term epilepsy, but when one considers how many of these children cease to have seizures before reaching adult life it is questionable whether a term with so many emotional overtones is justifiable.

Electroencephalography

Some help in this connection may be obtained from electroencephalography. Such specific abnormalities as a generalised spike and wave, spike and wave variants, generalised or focal spike discharges, can be regarded as ample support for prolonged anticonvulsant therapy. Less certain are the so-called slow wave dysrhythmias where the abnormality appears to be more in the nature of an immaturity of the record. As a child grows older, the amount of slow-wave activity of his electroencephalogram diminishes. Thus the normally theta-dominant record of a three-year-old would be distinctly abnormal if encountered in, say, a ten-year-old. Similarly, it is difficult to be certain how much slowing and increase in amplitude on over-breathing can be accepted as normal. It is probably wiser to regard such doubtful records as supporting a decision to use prolonged anticonvulsant therapy. It must, of course, be remembered that the diagnosis of epilepsy is not precluded by the finding of a normal electroencephalogram, nor is it established by an abnormal one.

It should be axiomatic, therefore, that all seizures should be prevented where possible, and an assessment of the likelihood of further seizures should be weighed against the slight inconvenience of treatment. In cases of doubt, my practice is to treat a child for two or three years on the assumption that such treatment is harmless and that it at once ensures, or attempts to

ensure, that the child has no more seizures and also remains free from the social disability in later life which a label of epilepsy unhappily confers.

After-care of Major Seizures

The decision when to initiate prolonged anticonvulsant care in children who have had major seizures is in the last resort empirical and indeed is closely related to the criteria of diagnosis of epilepsy. Where a direct cause has been found and dealt with, such as meningitis or tetany, there is normally no reason to suppose that any further seizures will occur. Yet even here one must bear in mind the possibility or even probability in some cases of convulsions themselves resulting in permanent cerebral damage with ensuing spontaneous seizures thereafter. Indeed there is a sense in which convulsions beget convulsions. One has only to see a protracted convulsion with its impaired respiration and cyanosis to appreciate that cerebral anoxia may be present, and the deterioration in an epileptic who has had a series of protracted convulsions is a common observation.

Drugs

Mention has already been made of the use of chloral hydrate for seizures in infants, where the dosage is restricted only by such sleepiness as keeps the baby from feeding normally. Where anticonvulsants are required for longer periods it is probably more convenient to substitute a drug that need not be given so often as chloral hydrate, which is normally given four- to six-hourly. In the first years of life phenobarbitone (15 mg.) gr. $\frac{1}{4}$ or primidone 0.125 g. may be given twice to three times a day. This can be increased in older children to twice this dosage, beyond which it is not as a rule useful to go. The addition of phenytoin 50 mg. daily in the first instance is often helpful, although doses of up to 100 mg. three

times a day may be needed in older children: too rapid an increase in dosage often produces ataxia. Rashes may result from any of these drugs but usually disappear on withdrawal of the drug and gradual re-introduction. 'Peganone' (or ethotoin, *N.N.D.* and *Martindale*), as a more easily tolerated hydantoin, is useful in doses up to 500 mg. three times a day, together with phenobarbitone or primidone where phenytoin has been unsuccessful or produced ataxia.

Ketogenic Diet and Fasting

The induction of ketosis either by fasting or diet or a combination of both has been known and recommended for over forty years without, however, gaining much popularity (Wilder 1921). Prolonged fasting, in which the child is encouraged to act and play normally with an intake of only water for three to four days, apparently has the disadvantage of hypoglycaemia added to the formidable practical difficulties. Without a preliminary fast it is difficult to achieve an adequate ketosis and the dietary imposition would be largely impractical outside an institution. Before the age of three years children are relatively resistant to ketosis (Williams *et al.* 1957). The subject is reviewed in detail by Bridge (1949).

Status Epilepticus

There is no precise dividing line between a prolonged generalised convulsion and status epilepticus. It is nevertheless true that the longer the convulsion, the more prolonged is the cerebral anoxia. Some of the children die and at autopsy there is frequently no obvious lesion of the brain to be seen. Others, especially after a prolonged unilateral convulsion, develop a spastic hemiplegia with a varying degree of residual mental defect and sometimes aphasia. In such cases although sedation must be prompt and sufficient it is equally important not to depress respiration to a

point where cerebral anoxia is further increased. The use of intramuscular phenytoin 50 to 100 mg. in addition to paraldehyde as described above is of great value.

Breath-holding Attacks

How far breath-holding attacks belong to the subject under discussion is perhaps a matter for argument. It is probably wise to distinguish between two types of attack. The classic breath-holding attack begins with a normally provoked expression of temper or fear: the child screams without pausing to breathe and eventually becomes cyanosed and unconscious. Immediately thereafter he recovers and the episode is over. Phenobarbitone appears to diminish the number of attacks but they usually cease in any case within a few months and do not recur. In the other type of attack the sequence of events is the same up to the point of unconsciousness, but after this the child has a major seizure. Here the breath-holding attack appears to provoke a major seizure and prolonged anti-convulsant treatment is wise, for it is not unusual for spontaneous major seizures to follow.

Minor Seizures

Various types of minor seizures may be encountered either alone or in association with major seizures. Whenever they occur they must be regarded seriously. Paradoxically a convulsion may be much less serious from the point of view of prognosis than a minor seizure, especially the lightning motor seizures of infancy. Lennox (1960) described the petit mal triad which includes 'true' petit mal, myoclonic and akinetic seizures. Other authorities, on clinical as well as encephalographic grounds, class the last two together as myoclonic (Gastaut 1954). True petit mal, with the classic momentary 'absence' or interruption of consciousness without any motor component (except perhaps a slight flickering of the eyes and momentary

pallor), is a rare condition even in childhood. It is almost always possible to demonstrate the classic 3 c/s spike and wave on electroencephalography (or more correctly, 4 c/s decreasing to 3 c/s in a prolonged burst). These seizures are resistant to the usual anticonvulsants and frequently become much worse when phenobarbitone or primidone is given. Most of them should respond to troxidone or paramethadione beginning with 150 mg. twice daily and increasing to 300 mg. thrice daily. Complications of this therapy are rare, although nephrosis and agranulocytosis have been reported. A newer compound, ethosuximide, has proved useful (250 mg. two to three times a day) in controlling the few that fail to respond to the diones.

It is in this group that photosensitivity is predominantly found both on EEG with photic stimulation and clinically when the history of seizures induced by gazing at the sky, at bright lights or even television is obtained. Self-induced seizures of this nature have been seen and present an interesting psychological problem (Hutchison *et al.* 1958).

Myoclonic Epilepsy

There is some confusion inherent in this term and Lennox would have us distinguish between myoclonus and myoclonic epilepsy (Lennox 1960). The term '*myoclonic*' is usually used to describe the brief motor seizures in which there is a generalised single muscular contraction, which in the infant is termed variously a 'salaam' fit, lightning fit, infantile spasm or ventral fit: in older children the characteristic is the sudden fall, usually forwards. The woe-begone picture of the toddler with several bruises on his forehead is a familiar picture to the paediatrician. These children do not so much fall as hit the ground and those hitherto called 'akinetic' probably belong to this category. The electro-

encephalogram frequently shows a spike and wave pattern although this is sometimes less clear than that found in 'true' petit mal, and all grades of abnormality can be seen from the spike and wave to the high amplitude delta activity with polyspike discharge which goes by the name of hypsarrhythmia. Something, perhaps, should be said about this term which is mentioned below in connection with myoclonic seizures and mental deterioration. The term was introduced by Gibbs *et al.* (1954) to indicate excessively high amplitude abnormality (*hyps* = high) in which random delta activity was accompanied by spike discharges of very high voltage. Although it is frequently seen in infants with myoclonic seizures or 'infantile spasms' it cannot be regarded as specific for any particular disease. Hypsarrhythmia appears to indicate a widespread centrencephalic disorder and may well differ from spike and wave variants (which are also seen in myoclonic seizures) in degree rather than kind.

Sudden Mental Deterioration and Hypsarrhythmia

The claims that ACTH has a beneficial effect on infants with 'infantile spasms' and hypsarrhythmia has recently focused attention on this condition: indeed, so firmly established is the concept of a pathological entity comprising myoclonic epilepsy, sudden mental deterioration and hypsarrhythmia, that a recent paper speaks of the 'treatment of hypsarrhythmia' (Trojaburg and Plum 1960). It cannot be too strongly emphasised that hypsarrhythmia is an electroencephalographic term for a non-specific disturbance and not a clinical diagnosis. It would seem wiser to adopt the attitude expressed in Illingworth's paper (1955), where he comments on the sudden mental deterioration of infants associated with seizures which were by no means always or

exclusively myoclonic. Moreover, the EEG findings in these children are not always those of hypsarrhythmia, nor is hypsarrhythmia always confined to those with myoclonic seizures. I have seen classical 'salaam' fits or infantile spasms with mental deterioration associated with a spike and wave abnormality of the EEG and unequivocal hypsarrhythmia in an infant with progressive mental deterioration and generalised convulsions but without myoclonic episodes.

A further important point commented on by Lennox (1960) and by Jeavons and Bower (1961) is the tendency for myoclonic seizures to cease as swiftly as they begin; they are seldom seen after the first few years of life. It is against this background that the claims of steroid therapy must be examined. The general view seems to be that ACTH gel given as 20 I.U. intramuscularly each day for about 20 days will be followed by cessation of the seizures in some third of cases. A higher figure of 11 out of 23 is reported by Jeavons and Bower (1961), who recommend four weeks' treatment. Although one might hope that early diagnosis and treatment might improve the prognosis there seems no evidence so far that the associated mental deficiency is in any way affected by treatment. All but one of my cases have shown evidence of severe mental retardation when first seen; this one child is perhaps worth mentioning in more detail. She presented at the age of nine months with a history of characteristic 'salaam' fits for the past three months. At this time she was already walking with support and her I.Q. was assessed at 110 on the Griffith's scale. Her EEG on admission (a sleep record) showed frequently repetitive generalised spike and polyspike discharges of high amplitude and she was given a 28-day course of ACTH gel 20 I.U. intramuscularly per day. Her seizures ceased within 4 days except for

two seen on the 14th day. An EEG during treatment showed less frequent spikes of lower amplitude while at the end of four weeks no further spiking was seen and her record was classed as normal. Time will tell whether her seizures will recur and it is of course possible that they would have stopped even without treatment. By the same token the fact that she is mentally normal cannot be necessarily attributed to the treatment; in fact, the history of three months' myoclonic seizures without any sign of mental defect is in itself unusual.

Anticonvulsants are usually disappointing in this group of seizures although a combination of primidone and acetazolamide has seemed effective in some cases and may be worth trying.

Our present understanding of this condition or group of conditions is still very incomplete but it would suggest that a course of steroids is worth trying in any infant who shows signs of sudden mental deterioration with coincident seizures of any type, whether or not the EEG shows all the features of hypsarrhythmia.

Behaviour Disorders in Epilepsy

The major problem of management of a child with seizures often lies in the behaviour rather than the fit. The association of behaviour disorders with epilepsy of rhinencephalic origin is well known, and the seizures may be major, minor or 'psychomotor', with various types of inappropriate behaviour as a manifestation of the seizure. While primidone has proved effective in many children as an anti-convulsant, its effect on the associated behaviour has been disappointing. There are, broadly speaking, two main patterns of abnormal behaviour seen in epileptic children. The one whose behaviour is reasonable except for unprovoked explosions of temper and aggressiveness: the other, the hyperkinetic syndrome, in which the child restlessly perseverates in all he

does, never remaining still for a moment and showing an insatiable curiosity which tires the moment it is aroused. Sedatives are ineffective and sometimes seem to make the patient worse. In my hands tranquillisers too have none of the magic inherent more perhaps in the name than in the drug, and only dexamphetamine, which is well tolerated, seem occasionally to be of some value. Mental retardation is often a feature in these cases and the more defective the child, the more hopeless is the treatment. Many an epileptic ends his days in an institution more because of his difficult behaviour than because of his mental retardation or epilepsy.

Long-term Care

Ideally, consideration of long-term care should be confined to the consideration of how long to maintain treatment in a child whose seizures are controlled. This is in any event a question to which only an arbitrary answer can be given. It is sometimes said that the diagnosis of epilepsy implies life-long treatment. On the other hand, if the view is correct that the tendency to seizures diminishes as the child grows, it might seem logical to omit treatment. It is not possible to lay down a hard and fast rule about this. The number of seizures and their duration, the presence of a family history and the electroencephalogram must all play a part in arriving at a decision. It is probably safe in a child who has had seizures for only a year or two, to omit treatment after three years of freedom. In some, however, the return of the seizures will require treatment once more.

Difficulties in schooling arise and these are not always directly attributable to the child's seizures. Obviously where mental

deficiency has also to be considered, the appropriate special school or occupational centre is unexceptionable. When an epileptic child has an intelligence within the normal range there should be no bar to his normal schooling provided that his seizures are kept down to a reasonable minimum with therapy. There are two factors which tend to interfere with this admirable arrangement: firstly there is the difficult behaviour of some epileptic children, and secondly there is the undeniable prejudice of some school-teachers against epileptic children. That this may readily become a vicious circle is too painfully obvious and the child who is singled out for unfavourable notice in a class often reacts with aggression and disobedience which results in his relegation to a special school. Special schools for the physically handicapped are ill-suited to epileptic children whose disability bears no relation to those of the other children at these schools and whose needs and capacity for education are not necessarily diminished by their handicap.

Where epileptics are also mentally defective their ultimate care in an institution raises problems little different from those of other defectives. In the mentally normal epileptic where the number and character of his seizures makes independent life impossible separate care in a colony is obviously desirable. Nevertheless, the behaviour disorders of some of these patients makes custodial care essential and it is obviously undesirable that both classes of patients should be treated in the same way or even in the same place. Hostels, sheltered employment, epileptic colonies and custodial institutions must all play a part in the long-term care of the epileptic.

SUMMARY

The rational management of seizures in infancy and early childhood can only depend upon as accurate a diagnosis as possible.

Symptomatic treatment with anticonvulsant drugs is required in all those who show a tendency to repeated seizures whatever the cause.

The use of ACTH in the treatment of children with sudden mental deterioration, seizures and hypersarrhythmia is described.

The behaviour disorders seen in some children with epilepsy often prove a more difficult social and therapeutic problem than their seizures.

The education and institutional care of epileptic children are discussed.

RÉSUMÉ

Le traitement des crises dans la première enfance et chez le jeune enfant

Le traitement rationnel des crises dans la première enfance et chez le jeune enfant dépend d'un diagnostic aussi précis que possible.

Un traitement symptomatique avec des médicaments anti-convulsivants est indiqué chez tous ceux qui ont tendance à répéter les crises quelle qu'en soit la cause.

L'emploi de l'ACTH est décrit dans le traitement d'enfants qui présentent une détérioration mentale soudaine, des crises, de l'hypsarrhythmie.

Les troubles du comportement observés chez certains enfants épileptiques représentent souvent un problème social et thérapeutique plus difficile à résoudre que celui de leurs crises.

L'auteur discute ensuite de l'éducation et du placement des enfants épileptiques.

ZUSAMMENFASSUNG

Die Behandlung der Anfälle im Säuglingsalter und in der frühen Kindheit

Die rationale Behandlung der Anfälle im Säuglingsalter und in der frühen Kindheit hängt von einer so genau wie möglichen Diagnose ab.

Symptomatische Therapie mit antikonvulsanten Mitteln ist angebracht bei allen denen die eine Tendenz zur Wiederholung der Anfälle, welche auch immer die Ursache sei, aufweisen.

Die Verwendung von AKTH für die Behandlung der Kinder mit plötzlicher geistiger Deteriorierung, Anfällen und Hypsarrhythmia wird beschrieben.

Die Betragungsstörungen die man bei einigen Kindern mit Epilepsie beobachtet, erweisen sich oft als ein schwereres soziales und therapeutisches Problem als ihre Anfälle.

Erziehung und Anstaltspflege der epileptischen Kinder werden besprochen.

REFERENCES

- Bridge, E. M. (1949) *Epilepsy and Convulsive Disorders in Childhood*. New York: McGraw-Hill Book Co.
- Gastaut, H. (1954) *The Epilepsies*. Springfield, Ill.: Thomas.
- Gibbs, F. A., Fleming, M. M., Gibbs, E. L. (1954) 'Diagnosis and prognosis of hypersarrhythmia and infantile spasms.' *Pediatrics*, 1, 66.
- Hutchison, J. H., Davidson, J. R., Stone, F. H. (1958) 'Photogenic epilepsy induced by the patient.' *Lancet*, 1, 243.
- Illingworth, R. S. (1955) 'Sudden mental deterioration with convulsions in infancy.' *Arch. Dis. Childh.*, 30, 529.
- Lennox, W. G. (1960) *Epilepsy and Related Disorders*. London: Churchill.
- Trojaburg, W., Plum, P. (1960) 'Treatment of hypersarrhythmia with ACTH.' *Acta Paed.*, 49, 572.
- Wegman, M. E. (1939) 'Factors influencing relation of convulsions to hypothermia.' *J. Pediat.*, 14, 190.
- Wilder, R. M. (1921) 'The effect of ketonuria on the course of epilepsy.' *Mayo Clin. Bull.*, 2, 307, quoted by Bridge (1949).
- Williams, M. L., Kaye, R., Kumagai, M. (1957) 'Studies on the mechanism of ketosis in infants and children.' *A.M.A.J. Dis. Child.*, 94, 499.

The Problems of Children with Cerebral Palsy in the West Indies

Illustrated by Two Case-Histories

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First Case: Delroy, a boy of 8 years, was brought up to the Children's Out-patient Department of the University College of the West Indies because of difficulty in walking and talking. His mother had had eclampsia at his birth, and for a month afterwards he had been blue and thought unlikely to live. He had in fact recovered, but his development had been slow. He sat up at one year, stood at 18 months, and started to walk at 2 years but at the age of 8 years he was only able to walk with difficulty. Talking was also greatly retarded. He did not say 'Mamma' until the age of 3 years, and at 8 years his vocabulary was limited and he had difficulty in articulation. However, he understood what was said, had control of his bowels and bladder, and was thought by his parents to be sensible. On examination he was rather small for his age but in good general physical condition. He would obey orders and was co-operative, and although psychiatric testing was not done it appeared that mentally he was not grossly subnormal. There was generalised spasticity of all four limbs, more marked in the legs than in the arms. Tendon jerks were exaggerated and the plantar responses extensor. His gait was unsteady, with marked scissoring. He walked on the tips of his toes; there was no ataxia. His speech was indistinct. In summary he was a spastic with a speech defect of normal or only slightly subnormal intelligence. In England this boy would probably be seen at an outpatient department or perhaps admitted to an assessment centre for spastics. His walking would respond to physiotherapy and training. Speech therapy would almost certainly improve his speech and he would be able to receive schooling.

This boy exemplifies many of the problems faced by cerebral palsied children and their parents in the Caribbean today. He lives in the country about 70 miles from the teaching hospital to which his parents have brought him by bus after getting up at 3 a.m. His father is a labourer earning about £2 a week, on which he keeps his wife and five children. It is obviously quite impractical for him to attend the physiotherapy department as an outpatient. There is a local hospital in a town a few miles from the patient's home. The Government doctor there is interested in the boy and has referred him to the teaching hospital. But he has no physiotherapist at his hospital. The teaching hospital, with 35 beds for children of all ages including babies, cannot afford to keep the boy in for the months which would be necessary to obtain appreciable improvement from physiotherapy and speech therapy. Even if he could be admitted the boy would receive no schooling, and admission would involve almost complete separation from his parents owing to the high cost of visiting. There is a rehabilitation centre run by the Jamaican Government, predominantly for polio patients but admitting a few spastics. The centre is always overcrowded and understaffed, and it would be many months before he could be admitted. He would be separated from his home, and schooling would be difficult because of his speech defect.

So the problem is discussed with the parents and the aetiology is explained. Simple exercises are shown to the mother, the parents are asked to encourage the boy in walking and talking, and he goes home. If he had lived in Kingston he could have received physiotherapy and speech therapy as an outpatient. If his parents had had more money they could have arranged for him to stay in Kingston with his mother to receive a trial of treatment. It is to be noted that the parents are fond of the boy and have obviously looked after him well.

Second Case: Marlene, aged 10 years, was brought by her mother to the Children's Outpatient Department because she could not use her left arm. The mother had noticed that at the age of one year the child was not yet standing and had taken her to the Kingston Public Hospital. She was found to have a left hemiplegia and was treated in the physiotherapy department for a year. By this time the child was walking well and the mother ceased to attend. Although she was handicapped she was attending an ordinary school. On examination, Merlene was a well cared for intelligent girl in good general condition. She had a left-sided hemiplegia. The leg was only slightly affected, so that, although the muscle tone was increased and the plantar response was extensor, her gait was normal. The left upper limb, however, was wasted, shortened and useless. There was some muscular power in the limb and on further examination it was evident that all movements were present though limited. She kept the limb in the hemiplegic position with a dropped wrist. The child's mother said that the girl never used the arm at all. Marlene lives in Kingston and her mother can bring her up for physiotherapy, so she may improve considerably. Regular physiotherapy from an earlier age might have prevented much of the disability. One wonders why, when there was such a good result from physiotherapy to the leg in infancy the mother ceased to attend.

In England such a patient would be followed up by the almoner to enquire why she had stopped attending. In Jamaica no

almoner service is available and the congestion of departments makes it impossible to chase defaulters.

Local Conditions

The British Caribbean consists of the newly formed Federation of the West Indies together with the mainland territories of British Honduras in Central America and British Guiana in South America, which is geographically not in the Caribbean at all. The University College of the West Indies, with its medical school in Jamaica, serves the whole area. Medical and other problems of the area are basically similar. The total population is between 3 and 4 million. The islands are densely populated, while the mainland territories, apart from the coastal belt of British Guiana, are largely empty lands. Ethnically the largest group is African in origin, descended from slaves brought from West Africa in the 17th and 18th century, with considerable European admixture. In Trinidad and British Guiana about 40 per cent of the people are 'East Indians' descended from Indians who were recruited as indentured labour in the 19th century. There are small numbers of Europeans, Chinese and Syrians. Indigenous Amerindians are found only on the mainland; very few Caribs still survive in Dominica. Spastics are found in all racial groups.

Agriculture is the main occupation of the area. Sugar is the chief crop, bananas, cocoa and citrus are also grown. Large estates and small peasant holdings exist side by side. Oil is found in Trinidad, and bauxite in Jamaica and British Guiana. Industry is beginning in Jamaica, Trinidad and British Guiana and is being encouraged. Tourists contribute to the national income, particularly in Jamaica. Throughout the area most of the population is poor. This is probably the greatest single problem for the cerebral palsied. Unemployment is widespread and under-

employment more common still. The sugar industry employs a large number of people during the crop season (about six months in Jamaica), but outside this season many labourers spend long periods out of work.

Causes of Cerebral Palsy

The causes are much the same as in temperate countries. Many deliveries are still conducted by untrained 'nanas', but labour on the whole is easy so that the amount of cerebral palsy due to birth injury is less than might be expected. Rh incompatibility is rarer than in the U.K., since only about 7 per cent of the population are Rh-negative. Sequelae to kernicterus are more likely to be due to ABO incompatibility, which is not infrequent. The late results of purulent meningitis are unfortunately not rare; too many of these cases come up for treatment too late in the illness. Encephalitis, presumably viral in origin, is by no means uncommon in Jamaica and sometimes leaves permanent brain damage.

The types of cerebral palsy found are again similar to those found in the U.K. Spastic tetraplegia, usually with mental defect, is common. Hemiplegias and diplegias, both mild and severe, do occur. There are some athetoids but cerebellar ataxia is not common.

Treatment Facilities

Throughout the area treatment facilities for the child with cerebral palsy are inadequate. This is because of a shortage of trained staff—doctors, physiotherapists and teachers—and the lack of funds. It is not so much that Governments and private social agencies are disinterested in the welfare of the cerebral palsied as that there just is not enough money or trained personnel to go round. There are not even enough teachers to teach normal children.

In Jamaica the mild cases—such as hemiplegics with a normal I.Q. whose

mothers can bring them to hospital—can receive physiotherapy at the Kingston Public (Government) Hospital or at the University College (teaching) Hospital. They can be supplied with any necessary appliance and can receive orthopaedic treatment. On treatment many of these children do well. The chief difficulty with this group is that of transport. Children living outside Kingston cannot be brought up for regular treatment and even those resident in Kingston often find bus services difficult. Also the mother may not be able to afford time off work to take the child to hospital.

After an epidemic of poliomyelitis in 1954, the Government of Jamaica, aided by public subscription, set up a rehabilitation centre for patients. Since then there has not been any serious outbreak of polio, so there is room in the centre for a few cases of cerebral palsy. There they can receive both physiotherapy and schooling as inpatients. In addition, some children attend the centre daily, being collected from a central point in Kingston by bus. Many parents cannot manage to get the child to the collecting centre, and many more children could benefit from attendance at the centre if there were more space. The Red Cross has recently opened a residential school for handicapped children attached to the rehabilitation centre.

In Trinidad the Red Cross Convalescent Home does admit cases of cerebral palsy, who receive physiotherapy and some occupational therapy but no schooling. In addition, the Princess Elizabeth Home for Handicapped Children has 80 beds for children over four years, with a school and physiotherapy facilities for all sorts of handicapped children. But, as in Jamaica, many of the patients have had polio and there are not nearly enough places for all the cerebral palsied children who would benefit from treatment.

In Jamaica outside Kingston, and in the

other territories, the facilities for 'spastics' are even less. A school for physically handicapped children has recently been opened in British Honduras; one of its patients is a severely handicapped boy of 12 years with contractures of both legs who had spent years in the local infirmary believed to be mentally defective, but was now progressing under the devoted care of a Roman Catholic Sister trained in teaching handicapped children (this teacher also taught the blind, the deaf, the rheumatic and a mild 'spastic'). In British Guiana the Red Cross also runs a convalescent home, where a few children with cerebral palsy receive schooling and occupational therapy.

But throughout the area the majority of 'spastics' at present receive no treatment at

all. Those living at home will attend school if they are physically capable of getting to school—which is not easy for a cerebral palsied child in mountainous country. Many children develop serious skeletal deformities and contractures from lack of treatment. When they get older, some of these eke out precarious livings by begging from tourists and others. Many children are cared for in the local almshouses, where they are fed and clothed but receive virtually no treatment.

The attitude of parents towards their cerebral palsied children is generally helpful. Mothers accept the responsibility for their care and will persevere in teaching them to walk. In general they do not expect the State or any other authority to take over the child's management.

SUMMARY

The medical aspects of the management of cerebral palsy in the West Indies are essentially similar to those in temperate countries. The chief difficulties are economic. The islands are poor and there is a general shortage of trained personnel. Treatment facilities for handicapped children, including the cerebral palsied, are scanty, owing partly to the lack of money and partly to the scattered nature of the islands.

RÉSUMÉ

Les problèmes des spastiques aux Indes Occidentales

Les aspects médicaux du traitement de l'infirmité motrice cérébrale aux Indes occidentales sont essentiellement semblables à ceux qui se posent dans les pays à climat tempéré. Les principales difficultés sont d'ordre économique. Les îles sont pauvres et manquent partout de personnel qualifié. Les possibilités de traitement pour les enfants diminués, y compris les infirmes moteurs cérébraux, sont rares, en partie faute d'argent et à cause de la dispersion des îles.

ZUSAMMENFASSUNG

Die Probleme der Spastiker in Westindien

Die ärztlichen Probleme der Behandlung der Zerebrallähmung in Westindien sehen denen, die sich in Ländern mit gemäßigtem Klima stellen, wesentlich ähnlich. Die hauptsächlichsten Schwierigkeiten sind ökonomischer Art. Die Inseln sind arm und überall fehlt es an ausgebildetem Personal. Die Behandlungsmöglichkeiten für Kinder mit organischen Minderwertigkeiten, einbegriffen Zerebrallähmung, sind gering teilweise wegen des Geldmangels, teilweise wegen der Verstreuung der Inseln.

REPORTS AND SPECIAL ARTICLES

The Royal Edinburgh Hospital for Sick Children

THE Royal Edinburgh Hospital for Sick Children has, for 65 of its 100 years, been housed in the present imposing building of red stone which is sited across parkland from the medical school (Guthrie 1960). Although lacking the spaciousness of some of its contemporaries, it was, when built, 'one of the most perfect hospitals in the United Kingdom'. With the later additions of a fine seaside branch, an infant unit and a further annexe, the Hospital has now a total of 244 medical, surgical, E.N.T., psychiatric and convalescent beds of which 154 are in the main building, while most of the dwelling houses to the immediate rear have been acquired for staff accommodation and ancillary services.

The Hospital has always had a fine teaching record, and here the illustrious Dr. John Thomson advanced the frontiers of paediatrics by his many original observations and by his international text-book. A new era opened in 1931 when Dr. Charles McNeil was appointed to the first Chair of Child Health in the United Kingdom. He had for some time been physician in charge of one of the Hospital's medical units and also Lecturer in the Diseases of Children. Furthermore his appointment as Honorary Paediatrician to the Edinburgh Royal Maternity Hospital in 1927 was one of the first such appointments in the country. His emphasis on child health rather than on the old convention of diseases of children and his efforts to establish close liaison between obstetrician and paediatrician altered the course of paediatric teaching

and practice not only within his own city but far beyond. His teaching was based very largely on the Hospital's wards and on the Hospital's pathology department where Dr. Agnes Macgregor and her assistants have contributed so much to a better understanding of this subject. Since 1946 Professor Richard Ellis has increased Edinburgh's contribution to child welfare both at home and abroad not only by his hospital teaching but by his writing, research, travel and continuous interest in children of all races both in health and in sickness. Among the list of former surgeons are the names of Sir Harold Stiles and Sir John Fraser, while colleagues of the latter include the present senior surgeon, Mr. James Mason Brown, and Professors Ian Aird, Norman Dott and Andrew Wilkinson.

Undergraduate instruction has been improved by the recent transfer of the University Department to an immediately adjacent site where better offices, tutorial rooms, library, pathology museum and laboratory space are available. The existing curriculum (which is soon to undergo extensive changes), currently provides for the teaching of Child Health with Obstetrics and Gynaecology in the same ten-week term of final year. Contact with the students is first established, however, during their third year when they receive a number of lectures on the physiology of childhood and a series of clinical paediatric demonstrations illustrating applied physiology. In their main course on Child Health the

students not only attend the medical and surgical wards daily but, increasingly, the outpatient departments in order to obtain a more balanced view of paediatric problems. With the use of the paediatric units of the Edinburgh Northern Hospitals a reduction in the size of clinics to about 12 students has been effected, and within individual medical charges sub-division into sections of 4 is possible. Attention is given not only to the expressions of disease but also to techniques of examining infants and children and to case-taking, while, although resident accommodation for students is still unavailable, attendances on take-in days and during University vacations are encouraged. The undergraduates also have twenty lectures which deal with the diseases of children (including behaviour disorders) and a number of others which cover such subjects as immunisation, adoption, feeding and the health services; ten lectures on the newborn infant; attendances at a well-baby clinic; a series of films and demonstrations dealing with development, admission to hospital, therapy and accidental poisoning; while they also attend the pathology museum several times in small groups for instruction by the Hospital pathologist. (The Final Professional Examination includes a written paper and oral, and both medical and surgical clinicals in Child Health.)

Post-graduates have attended the Hospital in large numbers for more than fifty years and since the Second World War the Edinburgh Post-graduate Board for Medicine has arranged instruction of various kinds there in both paediatrics and paediatric surgery for those with a specialist interest as well as for general practitioners. Since 1946 even closer international links have been forged by the employment of post-graduates from all over the United Kingdom, Australia, Canada, New Zealand, South Africa, India, Singapore

and Malaya, the United States of America, Europe, the Middle East, Burma and Thailand as residents, clinical assistants and registrars. All who have so served have enjoyed the experience and profited by the friendship, teaching and opportunity. The Hospital also houses an important school of paediatric nursing and provides training facilities for psychiatric social workers, almoners and speech therapists.

Members of the University Department and of the Hospital staff are currently conducting research in such fields as accident prevention, burns and fluid therapy, cerebral palsy and other aspects of child neurology, diabetes mellitus, the infant of the diabetic mother and related problems of neonatal physiology, prematurity, the Scottish Paediatric Tumour Register, speech disorders, and tissue culture, while members of the present staff have written or edited important books dealing with disease in childhood, child health and development, the surgery of childhood, diseases of the ear, nose and throat of children, and paediatric pathology, while another edits the *British Journal of Plastic Surgery*.

All this activity may suggest that the hospital organism is robust and healthy, but a crippling marasmus, the result of financial starvation, is already obvious. In spite of a sympathetic and progressive Board of Management* the present hospital, even with its modern décor and limited extension now nearing completion, represents an anachronism in a day when the world recognises the need for the provision of special care for children in hospital and the value of children's medical centres. It would be invidious to list the inevitable shortcomings which stem directly from

* English readers are reminded that in Scotland the Boards of Management of teaching hospitals are subject to the authority of Regional Hospital Boards.

lack of money, but in almost every department from the clinics to the kitchen, at a time when the annual intake of in-patients has risen by 12 per cent and new out-patients by 17 per cent over ten years, the staffing and accommodation have not increased in proportion (indeed, in some important departments the staff establishment has been cut) and are inadequate for the standard of service required. The problem is neither new nor unique in this country, but in an Edinburgh pulsing with excitement over plans for the complete replacement of adult hospitals, financial considerations prevent the provision of those services urgently required for children, and the money for even a temporary building programme, regarded by a unanimous staff as vital to the Hospital's

survival, has not been forthcoming so far. Rebuilding is unlikely until the needs of the larger adult population have been met, and that may mean not within ten or possibly even fifteen years or more. Yet in spite of this apparent inability to provide modern hospital facilities the great traditions of service and teaching are maintained *within these limits* by an enthusiastic medical and nursing staff proud of their traditions and desperately anxious to re-establish the hospital's previous pre-eminence. Above the main entrance, carved in stone, is a silent commendation or reproach to all concerned with the future of children's hospitals—INASMUCH AS YE HAVE DONE IT UNTO ONE OF THE LEAST OF THESE, YE HAVE DONE IT UNTO ME.

J. W. FARQUHAR

REFERENCE

- Guthrie, D. (1960) *Royal Edinburgh Hospital for Sick Children, 1860-1960*, E. & S. Livingstone Ltd., Edinburgh and London.

Cerebral Palsy in Denmark

Dr. Erik Hansen's survey

Reviewed by ROSS G. MITCHELL, M.D., F.R.C.P.E.

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THE publication last year of Erik Hansen's monograph on Cerebral Palsy in Denmark¹ was an important event, for this is the first time that a careful survey has been made of cerebral palsy in an entire country. Such a study must necessarily differ in emphasis and involve different techniques from more intensive surveys in selected areas, because of the much larger number of cases. Hansen's survey, which was originally planned to cover the whole population of Denmark, had to be confined later to those aged between 2 and 30 years, but the total number of cases—2,621—was still far too large for him to make a special examination of every patient. The study was therefore based partly on hospital and insurance documents and on questionnaires, and less than 20 per cent of the patients were seen by Hansen personally. However, 1,813 patients were examined by physicians familiar with cerebral palsy, and in Denmark—a country far advanced in the field of cerebral palsy—this implies a high level of diagnostic accuracy. If even 50 per cent of the 808 patients not examined by experienced physicians were diagnosed correctly, the overall accuracy would be as high as 85 per cent, and this is a very conservative estimate. Ascertainment of cases was very thorough, being helped by the well-organised system of civil registration in Denmark and by the statutory obligation to notify permanent handicap to a central Insurance Court. Thus only 2 patients could not be traced and these were excluded from the total.

The monograph very properly starts

with a definition of the types of case included in the term 'cerebral palsy'—a necessary preliminary which is too often omitted. This is followed by a useful review of previous work in the field. The study itself is based on data concerning 2,621 patients with cerebral palsy, believed to be all the survivors of those born in the years 1925 to 1953. The prevalence of cerebral palsy in Denmark in the age-group 2 to 30 years was estimated as 1.32 per 1,000, and a similar rate was found in children of school age. This is considerably lower than the prevalence in children of school age in Britain, which is slightly more than 2.0 per 1,000. Since the Danish survey was very complete, these figures do suggest that cerebral palsy is less prevalent in Denmark than in the United Kingdom. The distribution of cases throughout the districts of Denmark is described in detail but is of little epidemiological interest, for it depended on the location of Institutes and Homes for handicapped patients and did not reflect the true geographical incidence.

The chapter relating to the frequency of cerebral palsy is at times confusing and does not always make a clear distinction between prevalence and incidence at birth. Thus Ingram² is said to have calculated birth incidence in Edinburgh as 1.99 per 1,000 births, whereas Ingram's figure actually referred to the prevalence of cerebral palsy at the time of his survey in the population born between 1938 and 1952. It always seems rather unrealistic to discuss the birth incidence of a condition

which may not be recognisable for a year or more after birth, and Hansen's calculations of birth incidence are quite misleading because, as his own results show, many of the deaths in the earlier years of the period studied were not ascertained.

The section on the causes of death of the 199 patients who had died after the diagnosis of cerebral palsy had been made is particularly interesting. More than a third of the deaths were recorded as due to pneumonia. In about a quarter, the patient died in an exceedingly poor general condition, with recurrent high fever without demonstrable cause, and the cause of death could only be recorded as 'cerebral hyperthermia'. This pattern of mortality would probably accord with the experience of most paediatricians in this country. The Danish figures appear to show that there is a greater mortality rate among older cerebral palsied patients than among the general population of the same age; this might indeed be expected, but it was not the view of some of the earlier workers in cerebral palsy, such as Winthrop Phelps.³ Hansen calculates the average life expectancy of cerebral palsied patients to be about 30 years, but points out that this figure must be accepted with very considerable reserve.

The data on mortality are followed by a short review of 513 patients in whom the original diagnosis of cerebral palsy was not confirmed. One section concerns retarded motor development in patients not mentally defective, an especially intriguing group in view of the current interest in the 'floppy infant'. Of 61 such patients, Hansen considers that 7 were covered by the term 'hypotonia infantilis benigna' and that the remainder might be regarded as 'atypical cases of cerebral palsy'. While many people would not agree with this, most would concur with his view that such children deserve the same careful observation, and if necessary the same manage-

ment, as slight cases of cerebral palsy.

A comprehensive review of previous work on the classification of cerebral palsy ends with a description of the scheme Hansen used in his study. This classification is a reasonable compromise which will be intelligible, if not wholly acceptable, to most other workers. The percentage distribution of the various types is broadly similar to that found in most field surveys in this country:

Spasticity	78.5%
Athetosis	9.3%
Ataxia-tremor	4.0%
Spasticity + athetosis ..	4.6%
Spasticity + ataxia ..	2.5%
Unknown or uncertain ..	1.1%

The cases are also classified into 6 grades according to the degree of motor handicap. A very high proportion of the cases (63.9 per cent) are in the first 2 grades which, according to the author, together correspond to 'slight cases' in other surveys. This is a much higher proportion of slight cases than most workers in other countries have found. The difference may be a true one or may be attributable, as the author suggests, to the very complete case-finding. It must be accepted with caution, however, because of the impossibility of maintaining consistent standards when many different people have made the assessments.

No systematic study of the intelligence of the patients was undertaken, but an approximate evaluation was made in most cases on the basis of the records available. Of the 2,621 patients, 47.7 per cent were estimated as having normal intelligence. This is a higher figure than in most other series, but is consistent with the high proportion of slightly handicapped cases, for there was a close relationship between the level of intelligence and the degree of motor handicap.

Perhaps the weakest part of this study

concerns the associated disabilities, because no special search was made for them and routine records of speech, hearing and visual defects are of doubtful value. Thus, speech disturbances were recorded in 50 per cent of the 1,813 patients examined by physicians familiar with cerebral palsy, but in only 28 per cent of the 808 patients examined by less experienced clinicians. Disturbances of hearing were recorded in 4.3 per cent of those examined by specialists and in only 2.1 per cent of the other group. This shows how easily hearing defects are overlooked, even by experts, unless special methods of detecting deafness are adopted, for the careful studies of Fisch⁴ and Mowat⁵ indicate that the true incidence of deafness in cerebral palsy is about 25 per cent. It is probable that Hansen's figure of 25.5 per cent for oculomotor disturbances is also an underestimate.

Epilepsy is much more likely to be noted in routine recording than defects of the special senses, and this is borne out by the results of the study, which are similar to those of other workers. Thus epilepsy was recorded in between a quarter and half of the patients with spastic tetraplegia or hemiplegia, and in 1 patient in 10 of these with spastic paraplegia. The overall incidence of epilepsy in the 2,621 patients was 22.2 per cent.

In discussing aetiology, Hansen shows that he is well aware of the unreliability of retrospective obstetric data, and confines his discussion mainly to facts such as maternal age, birth rank and so on, about which there could be little doubt. In the 1,193 cases in which the mother's age at the birth of the child was known, there was a highly significant preponderance of mothers aged 35 years and over. There was also a small but significant preponderance of first-born infants and of twins. The recognised association between cerebral diplegia and premature birth is again emphasised by Hansen's data. A feature of interest was

that a majority of the patients in the group 'spastic monoplegia + paraplegia' were born in the last quarter of the year; this was even more pronounced among prematures.

Analysis of the treatment received by this large group of patients shows clearly the gradual evolution of care in Denmark during the past 35 years. At the beginning of the period many patients received no active treatment at all. Thereafter there was a gradual increase in the proportion treated by active methods, at first mainly orthopaedic operations. Only in the more recently born groups did physiotherapy come to assume its present dominant place in treatment.

The chapter dealing with training and occupation is potentially the most interesting and important. It could usefully have been amplified by an account of the general social and educational structure in Denmark and by some background detail of the pattern of employment in the country. Hansen's assessment of the need for accommodation in schools and other institutions is of particular interest, and should prove most helpful to those planning for the future. He finds that there is especial need for kindergartens designed for children with cerebral palsy. As in other countries, the provision for mentally defective cerebral palsied patients in Denmark has been deficient until recently and there is a great need for special departments for these children. There is also still a need for schools for the more intelligent children with cerebral palsy.

The discussion of employment concerns the 1,127 patients over the age of 15 years. Of these patients, 649 (56 per cent) were not employed, 303 were occupied and 175 were in training. On the premise that half of those undergoing training would complete it, Hansen calculates that about 35 per cent of the adult patients would 'manage

socially'. This rather high proportion may be due to the large number of slightly handicapped patients, who would have a high rate of employability. The interpretation of these data on employment would have been easier if they had been analysed in terms of the potentially employable rather than in terms of all patients of employable age. Nevertheless they form a valuable addition to the scanty information we have about the lives of people with cerebral palsy after they leave school. Like others who have studied the problem, Hansen finds that there is pressing need for occupational training and workshops for adults with cerebral palsy. 'The environment with which adult cerebral palsy patients with a moderate to severe handicap have to cope is often very sad and empty. A number of them stay at home quite unoccupied. They are often, and often feel themselves, a burden on their surroundings.' Small wonder that so many young adults feel bitter about the sudden loss of interest in them which all too often replaces the solicitude of earlier years.

A very large survey such as this, encompassing a whole country, serves mainly to indicate broad trends within the

country, and it is a mistake to place too much emphasis on detail, for generally the accuracy of such a field survey is in inverse proportion to its size. One cannot help feeling that at times the author has been tempted to overrate the reliability of his information, and one may justifiably question the propriety of detailed analysis of data which are acknowledged to be unreliable. Results such as these are certainly of great interest and value to those working in the field, who are aware of the problems involved and can give proper weight to the findings in the light of the information supplied. Hansen is obviously fully cognizant of the difficulties and pitfalls of this type of survey and is careful to give the detailed particulars which enable other workers to evaluate his report. The danger is that less experienced readers, or those who do not read the monograph sufficiently carefully, will place undue reliance on the accuracy of the data and thus draw unjustifiable conclusions. This risk should not be allowed to detract from the undoubted importance of Hansen's contribution, which will be a valued source of reference to all interested in cerebral palsy.

REFERENCES

1. Hansen, E. (1960) Cerebral Palsy in Denmark. Copenhagen: Munksgaard.
2. Ingram, T. T. S. (1955) 'A study of cerebral palsy in the childhood population of Edinburgh.' *Arch. Dis. Childh.*, 30, 85.
3. Phelps, W. M. (1941) 'The rehabilitation of cerebral palsy.' *Sth. med. J.*, 34, 770.
4. Fisch, L. (1957) 'Hearing impairment and cerebral palsy.' *Speech*, 21, 43.
5. Mowat, J. (1961) In *Cerebral Palsy in Childhood and Adolescence*. Ed. Henderson, J. L. Edinburgh: Livingstone.

Electrophysiological and Biochemical Methods in Child Neurology

Annual Conference of Czechoslovak Paediatric Neurologists, Prague,
May 25 and 26, 1961.

Reported by Dr. IVAN LESNÝ (Prague)

The Importance of EEG in Child Neurology. Introductory Report.

By I. LESNÝ (Prague).

Electroencephalography as a method of investigation in child neurology is now reaching its peak importance. Although nowadays there are many criticisms of EEG, it has helped to elucidate the nature of epileptic processes, cortico-subcortical inter-relations and many kindred problems.

As a method of localising intracranial tumours, EEG has less value in children than in adults because expanding hemispherical lesions are rare in childhood; routine EEG examination is also of less importance for localising posterior fossa tumours. However, this examination plays a much greater role in the detection of epilepsies and paroxysmal diseases in children because of their greater prevalence in childhood. While the brain is still immature, there is a special liability to paroxysmal conditions, such as sleep-walking, nocturnal terrors and febrile convulsions, and in all these conditions EEG examination can indicate the relation of the symptoms to epilepsy, and thus help towards successful treatment.

In recent years the EEG has become increasingly important in the diagnosis of inflammatory, demyelinating and degenerative diseases of the central nervous system. In the encephalitides, especially the toxi-infectious group, there is the striking case of the clinically recovered child who yet has severe EEG abnormalities: the clinical symptoms do not indicate the underlying biological lesion. In such cases one must be careful not to give a good prognosis when the EEG shows abnormalities.

The demyelinating diseases, such as van Bogaert's subacute sclerosis, the degenerative diseases, like Friedreich's ataxia, head injuries and their sequelae—all have specific or semispecific EEG findings. The information obtained from EEG records must be carefully compared with the results of other tests; EEG observations alone are not sufficient for a diagnosis, which ultimately rests with the clinician.

The Application of Induced Rhythms and Evoked Potentials in Children.

By A. HRBEK (Prague).

The method of photo-induced rhythms and evoked potentials was used in the investigation of 70 children of 5–17 years, and 40 newborn prematures. The results varied according to whether the children were over or under 10 years. Visually evoked potentials were

recorded in prematures during the first few days of life. These potentials were distinguished by their morphology, greater variability and longer latency (192 m. sec.). Non-specific responses were present in only 25 per cent of the children and these responses develop in the later weeks of life.

EEG in Newborn Infants with Perinatal Injury to the Central Nervous System.

By I. BREŽNÝ and Š. SRŠEŇ (Košice).

Twenty newborn infants with signs of perinatal brain injury were investigated by EEG in the first few days of life. The babies were divided into four groups according to their clinical symptoms. It has been supposed that EEG investigations may be useful in diagnosing perinatal brain injury in the first few days of life, but the correlation between clinical and EEG findings is not always perfect. Sharp waves are frequently seen in the EEG of the newborn but their significance is not always clear. As a rule manifestations of convulsive disorder are focal. About half the babies with intention tremor and reflex tremor showed in the EEG a remarkable high-amplitude rhythmical activity with a frequency of 7-9 c.p.s. in bursts and runs.

EEG Changes in Cerebellar Hypogenesis.

By I. LESNÝ (Prague).

EEG records were taken in 10 children aged 1½ to 8 years in whom the diagnosis of cerebellar hypogenesis had been made by air-encephalography. Seven of the children had bilateral rhythmic and synchronous slow waves 3-4½ per sec. in the occipital or occipitotemporal regions. Of the remaining 3 children, 2 had signs of delayed development of the potentials, one of them had focal spikes, and one had a dominant fast 20 per sec. activity.

Synchronous slow waves in the occipital region are a not unusual finding in degenerative diseases or in tumours of the cerebellum. They are a typical EEG feature in cerebellar hypogenesis. In the 3 patients whose EEG abnormality was of a different type, the changes were probably caused by other associated developmental anomalies.

EEG in Children with Cranial and Cerebral Trauma.

By R. VOJÍŘ, J. GUTWIRTH and F. FOJTÍK (Prague).

The authors reviewed the EEGs of 50 children with cranial and cerebral injuries of varying degree, the series comprising 28 boys and 22 girls, aged 6 months to 15 years. Practical problems in the use of the EEG, and its importance in the clinical evaluation of the severity of injury, in treatment, in prognosis and even for solving possible forensic questions were studied, and the following conclusions were reached. The normal EEG confirms the clinical diagnosis of concussion when the accepted clinical criteria are coma, lasting not longer than 15 minutes, and the absence of neurological findings. Contusion might be suggested when there are clinical signs of only slight injury, if the EEG shows abnormalities which do not disappear within a year. A slight contusion might be suggested in cases where the head-injury, clinically accepted as slight, is followed by rapidly disappearing EEG abnormalities. Contusion with depressed electrical activity suggests additional haemorrhage.

Photo-stimulation is sometimes useful for provoking pathological activity and for giving information on the dynamics of cortical rhythms, which seem to be affected first in head injuries.

The EEG in Children with Expanding Intracranial Lesions.

By J. STEIN and M. ZUKLÍNOVÁ (Prague).

The EEG records of 61 children with expanding intracranial lesions have been examined. The patients' ages ranged from 1 to 15 years. They included 36 with lesions in the posterior fossa, 15 with hemisphere and 8 with suprasellar lesions; another 2 children had multiple tumours scattered throughout the brain. The EEG records of supratentorial expanding processes were invariably abnormal; only 3 normal EEGs were found in the infratentorial cases, and these all had an intraparenchymatous tumour in the medial part. The most frequent pathological finding was slow (delta) waves which tended to be localised to the occipital region. Suprasellar processes more often showed diffuse changes, chiefly episodic ones. In the cases of expanding lesions in the posterior fossa, the focal changes more frequently appeared over the opposite brain hemisphere, but even in these cases the changes were mostly occipital. The authors try to distinguish between focal occipital EEG changes evoked by tumours actually located in this region and remote focal changes caused by posterior fossa tumours.

The authors then compare their findings in children with the results they have obtained in EEG examinations of 419 adults with expanding intracranial lesions. They formulate basic rules for detecting in an EEG record the presence of an expanding lesion as well as its localisation. They emphasise, however, that the EEG alone is not usually sufficient for the diagnosis of an intracranial tumour. The EEG findings must be correlated with the clinical ones, and the diagnosis has usually to be confirmed by contrast X-ray examination.

The EEG in Children with Enuresis.

By V. KOLÁŘ, L. SYROVÁ and J. GUTWIRTH (Prague).

The EEG findings in 50 enuretic children, aged 4 to 15 years, showed no signs of organic lesions. Children with mental defect, serious behaviour disorders or epilepsy were excluded. Of the 50 children, 36 had been continuously enuretic since birth; 14 had had periods of a year or longer when micturition had been fully controlled after the age of three. The EEG abnormalities found had no specific character. There were no spike-and-wave complexes, no spike activity and no focal changes. In the group of children enuretic since infancy far more EEG abnormalities were discovered (68 per cent) than in the other group, of which only 14 per cent showed abnormalities. The EEG may help to distinguish among the cases of so-called essential enuresis the existence of a mild encephalopathy, whereas other cases are predominantly psychogenic. These conclusions support other findings which show that epilepsy has only a minor connection, if any, with enuresis.

EEG Abnormalities in Obstetric Brachial Paralysis.

By I. LESNÝ (Prague).

Obstetric brachial paralysis, being a lower motor nerve lesion, has not so far been subjected to EEG examination. It is a surprising fact that in 4 such cases severe EEG abnormality was detected. These children had no other evidence of a c.n.s. lesion. Syn-

chronous episodic slow waves (2-3 per sec.) activity was seen. This slow wave activity had an occipital predominance and in 2 out of 4 cases was contralateral to the paralysis. In the remaining 2 it was bilateral.

The brachial paralysis being caused by an obstetric injury in all 4 cases, the EEG abnormality must be ascribed to subclinical brain damage caused by the same factor. We cannot say whether the lateralisation of the abnormality in two of our cases was caused by insufficient afferent impulses from the paralysed limb.

The EEG and Behaviour Signs in Sleeping Infants.

By M. RÖSSLER and D. DITRICHOVÁ (*Prague*).

In 8 infants the authors describe EEG changes in various phases of sleep and their relation to the frequency of respiration, to motor activity and to the position of the eyelids.

Electromyographic Studies in Children with Signs of Spasmophilic Neuropathy.

By Z. KUNCOVÁ, V. ŠKORPIL and J. KREDBA (*Prague*).

The authors tried to confirm the clinical diagnosis of spasmophilia in 20 children aged 13-14 years, without signs of tetany and with normal blood-calcium levels, by means of natural and stimulated electromyography. Ischaemia lasted 10 minutes and hyperventilation 5 minutes. They compared their findings with those in 11 healthy children of the same age. In 17 patients there was an increase in muscular reactions during the ischaemia.

The authors conclude that electromyography is of great value in the diagnosis of spasmophilic neuropathy.

Electromyography in Diseases of the Nervous System in Children.

By B. DRECHSLER (*Prague*).

A concept of the electrophysiology of muscle contraction is given. Electromyograms—i.e., recorded action potentials of the motor units—can be obtained during muscular contraction by inserting needle electrodes. In this study the value of the technique of 'stimulation-detection' was assessed and illustrated by specific examples. Cases were included to indicate how electromyographic data can be used in the diagnosis of traumatic lesions, such as paralysis of the facial nerve or brachial plexus. Electromyography can be used in the differential diagnosis of muscular hypotonias in childhood—for example, muscular dystrophy, Oppenheim's amyotonia, spinal progressive muscular dystrophy, muscular hypotonia related to upper neuron motor lesions, and other structures of the central nervous system. The value of determining the conduction velocity of peripheral nerves is illustrated by specific examples.

An Electromyographic Study of Dermatomyositis and Polymyositis.

By B. DRECHSLER and J. VACEK (*Prague*).

This report consists of the EMG study of the first 10 patients observed in a general study of dermatomyositis and polymyositis in children. The diagnosis was made on clinical and laboratory findings and biopsy. The following typical patterns of electrical activity were found: (1) Resting activity in the form of fibrillation potentials or motor unit potentials with low frequency (6 per sec.). It was difficult to obtain a resting state. (2) During voluntary

contraction the pattern of gradation of muscle contraction was disturbed. (3) Electrical activity was elicited more easily by rapid passive motion in myositic than in normal muscle. (4) On the insertion of the needle electrode, long-lasting trains of potentials and in two cases myotonic discharges occurred. (5) The mean action potential duration was generally diminished. (6) About 20 per cent of the potentials are polyphasic, compared with only 3-5 per cent in normals. (7) There is a relationship between the EMG findings and the biopsy results in the myositic muscles.

Use of Elastic Splints in Spastic Hemiparesis.

By J. PFEIFFER, K. OBRDA and J. KRAUS (Prague).

The authors describe a splint made of elastic material, which they used for the correct positioning of affected limbs, especially the hand. Electromyographic examination of muscle co-ordination, using plain needle registration, showed that after the application of the splint spontaneous resting potentials disappeared. This shows that the action potentials of the flexors of the fingers when splinted exceeded their electrical activity before splinting.

Clinical Application of Kymography in Extrapyrimal Disease of Children, especially Chorea Minor.

By Z. MAJEWSKA and K. SZELOZYŃSKA (Gdansk, Poland).

A kymographic study was made of 20 children aged 5 to 13 years and a control group. The child was made to hit a rubber ball repeatedly against a hard base. The ball was connected with a feather through a Marey tympanum. Each stroke was registered on the tape of the kymograph as a steep rising line and a less steep descending one. The amplitude of each stroke was the same and the strokes were made rhythmically. A study was first made with one hand and then with the other. Preliminary study for dominance showed that it made no difference which hand was used first: the graph obtained from each hand was the same.

Children with chorea minor showed big changes in form, amplitude and rhythm. The investigation established that although chorea minor is generally regarded as a symmetrical disease, in only 3 out of 20 cases were the kymographic changes similar in both hands. In 17 cases the changes were more obvious in one hand, usually the right (10 cases). Even in the 3 cases in which hemichorea was diagnosed, the kymographic investigation showed bilateral changes.

Kymography also showed the existence of paresis by a reduction in amplitude on one side. Frequently kymography showed the continuation of small changes in spite of the regression of clinical symptoms, and therefore gave some indication of when treatment could be ended.

Regulation Mechanisms of Carbohydrate Metabolism in Stress Reactions in Children.

By V. HRAZDIROVÁ (Brno).

Fluctuations in blood-sugar level were studied in neurologically ill children after stress acting either directly on the c.n.s. or at the periphery. Air insufflation at pneumoencephalography represented the central stress factor and the application of cold to the limbs the peripheral stress. Results were compared with the fluctuations in blood-sugar level noted

after the application of adrenaline and dextrose. The timing of the fluctuations in blood-sugar level was the same after central stimulation, after intramuscular adrenaline, and after oral administration of dextrose. Only small and irregular fluctuations, never outside the normal range, were seen after peripheral stress.

Penicillamine as a Diagnostic Aid in Wilson's Disease.

By B. BLEHOVÁ and L. HEYROVSKÝ (Prague).

In typical cases of hepato-lenticular degeneration the diagnosis is easy, but sometimes, especially in the hepatic form, it is difficult or even impossible. The biochemical methods of diagnosis are summarised here, and the use of penicillamine is suggested as a diagnostic aid. In 3 patients with Wilson's disease, the copper excretion rose from 800 to 3,000 gamma in 24 hours after 1 gramme of penicillamine, while in 7 healthy controls the excretion did not exceed 162 gamma in 24 hours and in 4 patients with hepatic cirrhosis the maximal rise was 180 gamma.

Some Neuro-radiological Observations in Childhood.

By D. MÜLLER (Berlin).

Brain lesions can be diagnosed and the time of their appearance determined by studying X-ray films of the skull sutures shortly before and after birth. The author describes the pressures on the skull which are changed by exogenous factors, such as rickets. Knowledge of the normal pressures exerted on the growing brain is necessary to differentiate endogenous from exogenous causes of deformity. Even in a static technique like radiology, a knowledge of the principles of development can therefore be useful.

OBITUARIES

Arnold Gesell

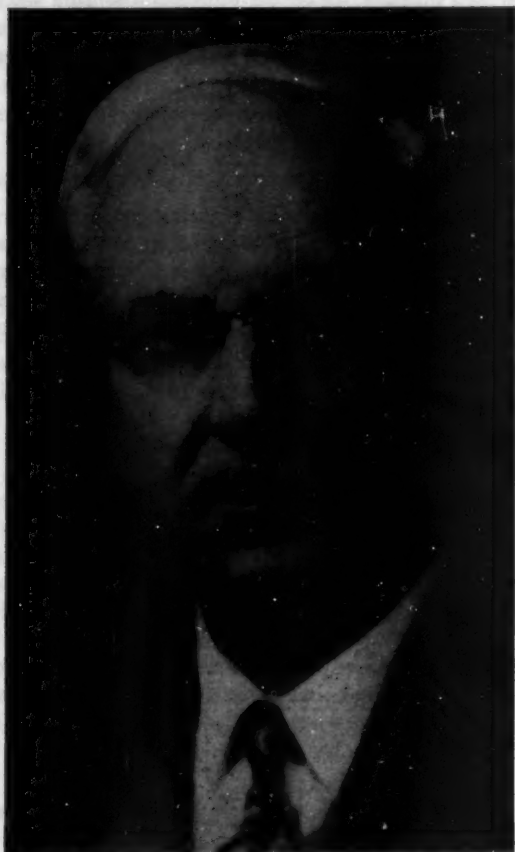
DR. GESELL, who died in May this year, devoted his whole life to the task of acquiring comprehensive and detailed knowledge of the behavioural development of the normal child. He was a pioneer in the field of developmental psychology to which he made a striking and original contribution, and has earned a wide public through his many writings and publications.

Born in Wisconsin in 1880, Dr. Gesell graduated from the University of Wisconsin in 1903. He taught at the Los Angeles State Normal School from 1908 to 1910, became professor of education at Yale, where he later graduated in medicine, and in 1915 was appointed to the Chair of Child Hygiene at Yale. From 1928 to 1948 he

was attending paediatrician at Newhaven Hospital. After 1948 he retired from academic posts and became research associate with the Harvard Paediatric Study for four years. He became president of the American Academy for Cerebral Palsy, and was a member of the National Research Council from 1937 to 1940.

In 1911 Gesell established the Yale Clinic of Child Development, and during the next decade, at a time when interest was centred predominantly on the problems of intellectual retardation and concerned itself necessarily with older children, began his systematic

studies of normal pre-school children, starting with two-year-olds and finally covering a span from birth to



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five years. Following Binet's work, Gesell aimed to describe the full range of normal behaviour in children. His method of examination was exacting and thorough; experimenting with everyday objects (a ring on a string, coloured bricks, etc.) he determined what sort of activity might be expected of the average infant when protected from outside interference. For these purposes Gesell used a one-way screen, through which the examination made by himself or another member of the team was watched by the rest of the team, and running commentaries made during the examination were recorded for later discussion. Gesell also made use of photographic and cinematic demonstration, so that his methods could be duplicated.

Gesell organised his research in terms of four major fields of behaviour, comprising: (i) motor characteristics, including posture and skills; (ii) adaptive behaviour, including perceptual, manual and verbal adjustments; (iii) language experience; and (iv) personal and social adjustments—to domestic life, the impact of culture, etc. On the basis of his studies he devised a *schema* of examination at 6-weekly intervals in the first year of life, which he claimed was predictive. By and large the *schema* does apply to severely subnormal children though less so for the normal range of intelligence. More recently, it is

true, Gesell's studies have been criticised for the small number of children studied on which his norms were based. But his contribution remains unchallenged; he was a pioneer in his effort to find criteria for the early detection of subnormality and to understand the developmental history of the normal child, and all paediatricians acknowledge their indebtedness to him. Gesell's orientation was in noticeable contrast to psycho-analytical theory, to which he was antagonistic; he was concerned with behavioural aspects of development and aimed to put the study of the normal infant on a scientific basis. In this he was first in the field.

One has only to read through the vast list of his publications to realise how wide were his interests and how comprehensive his enquiries. Gesell studied the developmental history of the newborn up to two weeks of life, and he carried out a study on groups of prematures. He found time, while still a consulting paediatrician, to study twins from infancy to adolescence, the development of vision, the feeding behaviour of infants, the fears of childhood and many other subjects relating to his field. He applied his work to the problems of selecting for adoption and to prediction in all its aspects. It is a tribute to his work that so many psychologists and doctors came to work and study under him.

RUTH LEYS

LETTERS TO THE EDITOR

The Blood-Brain Barrier

SIR—Dr. Benson and Dr. Joseph, in their letter in the October *Bulletin*¹, take me severely to task for what they conceive to be my views⁴ on the pathogenesis of kernicterus, and particularly on the value of prophylactic exchange transfusion in suitable cases of haemolytic disease of the newborn. It is presumably a measure of my failure to express myself clearly that they should have been led into attributing views to me that I do not hold. They saddle me with views which they admit I did not state and then attack them for being dangerous. They also consider that a number of my statements are refuted by observations which I do not cite. If they had read my original review³ (of which my annotation was only a summary), and those of my papers which are referred to therein, they would have found many of their objections answered.

In order to remove all doubt, let me state unequivocally that the evidence for the usefulness of prophylactic replacement transfusion for averting kernicterus in suitable cases of haemolytic disease of the newborn appears to me overwhelming. I know of no contrary opinions and have certainly never held any myself. It by no means follows, however, that the same manoeuvre averts kernicterus in other circumstances. There is room for doubt whether it is useful in the non-erythroblastotic jaundice of premature babies, for example, and there is still considerable discussion on this point.^{6, 7} My remark that kernicterus 'appears to be averted by prophylactic replacement transfusion' clearly applied to kernicterus as a whole, and in no way implied that I am unconvinced by the evidence for its usefulness in haemolytic disease. Neither am I advocating that it should be dropped at this stage even in the prematures. I am only putting forward a hypothesis.

The old 'primary damage' hypothesis—that damage to the brain must occur *before* bile pigment or any other protein-bound foreign substance can enter the tissue—is still, in my opinion, a better explanation of the known facts than the newer hypothesis which is now almost universally held. For those who formulated the original hypothesis, the already damaged brain was only incidentally stained by the bile pigment. However, it is now certain that the pigment will produce still further damage after it has entered. Contact with bile ruins even dead tissue for histology, and there is good evidence that it interferes with oxidative phosphorylation of brain mitochondria.⁵ (The reference¹⁵ quoted by Dr. Benson and Dr. Joseph for this is, incidentally, not concerned with brain. It is solely concerned with isolated liver mitochondria.)

The most important step is penetration, and I believe it to be most unlikely that protein-bound bilirubin would enter a normal brain at any age. It is quite certain, however, that any kind of lesion in the brain will allow the pigment to enter it, even in adults, and since the distribution of stained areas in kernicterus so closely resembles that of 'anoxic' and other metabolic lesions, it does not seem to me unreasonable to continue to postulate 'primary' (?anoxic) damage' in the pathogenesis of kernicterus.

Dr. Benson and Dr. Joseph ask me to explain why many 'babies with hyperbilirubinaemia, who have had no evidence of anoxia, have developed kernicterus.' I did not anticipate having to spell out the complexity and sophistication of current views on anoxia. The

expression 'no evidence of anoxia' is utterly worthless in this context. Throughout this discussion, it is vital to remember that permanent, irreversible brain lesions can occur after *transient* insult. Anoxia in its widest sense need only operate once for a few minutes for permanent brain lesions to be produced in babies who subsequently may show 'no evidence of anoxia.'

I am then asked 'why kernicterus can be prevented by exchange transfusion.' Of course I do not know. But Dr. Benson and Dr. Joseph take altogether too simple a view in implying that reduction of circulating bile is its only effect, particularly in haemolytic disease.

Another point raised was the well known relationship between the level of circulating bilirubin and the incidence of kernicterus. Here again there are many alternative explanations, and I would have thought it axiomatic that a relationship, however close (and this one is not always very close), was not inevitably a causal one. Supposing, for example, the same aetiological factor (?anoxia in its widest sense) were responsible for both the brain damage and the hyperbilirubinaemia? Could it not then follow that the more pronounced this hypothetical factor in the aetiology, the more likely would the child be to develop kernicterus, and the higher would be its bilirubin?

Dr. Benson and Dr. Joseph doubt my contention that 'it is no longer believed that there are differences between adult and neonatal brains in their relationships to blood-borne dyes,' and quote the reservations of Miller and Hess¹⁰ that it may be 'that the barrier is less complete in the young animal and that its permeability depends to some extent upon the substances used to test it.' By this reservation Miller and Hess mean that the passage of a substance from blood to brain depends, obviously enough, on the substance under consideration. Neurochemists have known for a long time that many normal metabolic substances, such as phosphate and cholesterol or its precursors, pass more rapidly into the immature brain during the period of myelination than subsequently. There is a growing reluctance, however, to attribute this type of phenomenon to a barrier permeability restriction. It is much more likely to be a reflection of the changing metabolic requirements of the brain as it grows and incorporates these very substances into its structure.^{2,3} The changing blood-brain relationships of these normal brain constituents at different stages of development are in marked contrast to the constant behaviour of protein-bound dyes which do not appear to enter the brain at any age, *unless the brain is previously damaged*. I regard Miller and Hess¹⁰ as the key reference here, in spite of their reservations about metabolic substances. This point defies concise summary, although I tried to present it in the fourth paragraph of my editorial. It is very fully discussed elsewhere.³

The literature contains many accounts of attempts to produce kernicterus-like lesions experimentally by the injection of bilirubin into newborn kittens, puppies and rabbits^{9, 11} and of its occurrence in Gunn rats.⁸ Close scrutiny of these results, however, nearly always reveals that generalised abnormalities are present in these animals such as may quite well lead to brain damage, which may, therefore, not be *directly* attributable to the bilirubin. Rodzilsky¹¹, for instance, found that preceding brain damage was required in newborn puppies and rabbits to obtain nerve-cell staining, and that even in his kittens 'haemorrhagic and exudative lesions 'gastroenteritis' and damage to the liver and kidneys were produced simultaneously (*sic*) in the newborn animals'. The same author¹² states, 'Furthermore, it is not always sufficiently stressed that the lesions of neonatal hyperbilirubinaemia are by no means restricted to the central nervous system. Among other organs

and systems shown to be damaged in cases of neonatal jaundice with kernicterus are the endothelia of the capillaries and serous membranes, the gastrointestinal tract, the liver (*sic*) and the kidneys.' It is a little hard to have this author quoted so selectively against my point of view, when, as I have demonstrated, it is quite possible to support my own opinion by equally selective quotations. Complementary with this, Silverman¹⁴ picturesquely adds, 'I should like to remind other blind men holding various other parts of the bilirubin elephant that there are competing causes of brain damage in premature infants; bilirubin injury is not the only or even the most common cause.' It only needs to be added that anyone who has seen Gunn rats knows them to be very peculiar animals indeed. It is wrong to imply that jaundice is their only defect. To summarise this part of the argument, no experimental kernicterus has been produced that I know of without there being concomitant sickness elsewhere in the animal. To speak of it having been produced in healthy, newborn kittens¹¹ has misled Dr. Benson and Dr. Joseph in a way that could have been avoided by closer scrutiny of the original paper.¹³

It is with great reluctance that I have written in such an argumentative manner. There is disturbing evidence in many quarters of a widening rift of misunderstanding and prejudice between research workers and clinicians, and it is up to both sides to be eternally vigilant to prevent this. But as long as critics invent things to criticise, when there is, after all, no shortage of disputable points to discuss, there will be no progress.—Yours, etc.

Department of Physiology,

JOHN DOBBING

London Hospital Medical College, London, E.1.

REFERENCES

1. Benson, P. F., Joseph, M. C. (1961) 'The blood-brain barrier.' *Cer. Pal. Bull.*, 3, 510.
2. Davison, A. N., Dobbing, J. (1961) 'Metabolic stability of body constituents.' *Nature, Lond.*, 191, 844.
3. Dobbing, J. (1961) 'The blood-brain barrier' *Physiol. Rev.*, 41, 130.
4. — (1961) 'The blood-brain barrier' *Cer. Pal. Bull.*, 3, 311.
5. Ernster, L., Herlin, L., Zetterström, R. (1957) 'Experimental studies on the pathogenesis of kernicterus.' *Pediatrics*, 20, 647.
6. Freedman, A. M. (1961) 'The neurological sequelae of hyperbilirubinemia of prematurity.' In *Kernicterus*, Toronto: Univ. of Toronto Press, p. 20.
7. Grewar, D. A. I. (1961) 'Experiences with kernicterus in premature infants.' *Ibid.*, p.13.
8. Johnson, L., Sarmiento, F., Blanc, W. A., Day, R. (1959) 'Kernicterus in rats with an inherited deficiency of glucuronyl transferase.' *Amer. J. Dis. Child.*, 97, 591.
9. Kuster, F., Krings, H. (1950) 'Blood destruction and cerebral damage in haemolytic disease of the newborn.' *Lancet*, i, 979.
10. Miller, J. W., Hess, A. (1958) 'The blood-brain barrier: an experimental study with vital dyes.' *Brain*, 81, 248.
11. Rozdilsky, B. (1961) 'Experimental studies on the toxicity of bilirubin.' In *Kernicterus*, Toronto: Univ. of Toronto Press, p. 161.
12. — (1961) 'Toxicity of bilirubin in adult animals' *Arch. Path.*, 72, 8.
13. — Olszewski, J. (1961) 'Experimental study of the toxicity of bilirubin in newborn animals.' *J. Neuropath. exp. Neurol.*, 20, 193.
14. Silverman, W. A. (1961) 'The effects on premature infants of sulfisoxazole in the neonatal period.' In *Kernicterus*, Toronto: Univ. of Toronto Press, p. 196.
15. Zetterström, R., Ernster, L. (1956) 'Bilirubin, uncoupler of oxidative phosphorylation in isolated mitochondria.' *Nature, Lond.*, 178, 1335.

Dr. Dobbing's letter has been seen by Dr. Benson and Dr. Joseph who write as follows:

SIR—We are grateful to Dr. Dobbing for his clearly expressed statement that in suitable cases of haemolytic disease of the newborn replacement transfusion averts kernicterus.—Yours, etc.

Guy's Hospital,
London, S.E.1.

PHILIP F. BENSON
MICHAEL C. JOSEPH

Is Treatment Really Necessary?

SIR—The question of the value of 'treatment' for children with cerebral palsy, as against parent guidance and good home management only, has recently been occupying the thoughts of a large number of specialists in this field. The main argument against treatment appears to be based not only on the lack of any estimate comparing the effect of one kind of treatment with that of another—e.g., of a neurological/developmental approach with that of an orthopaedic one—but also on the absence of any statistically conclusive evidence of the value of any kind of treatment.

Some hold the view that treatment is of little or no value and that cases which improve with treatment might have progressed equally well without it. One might reply to this argument that it has no proven facts to support it, and that no one can say with certainty that *every* child will improve without treatment. It seems surprising that the effect of treatment is questioned or denied in every case of cerebral palsy. In view of the great variety of conditions all grouped under the term 'cerebral palsy', it is unlikely that treatment is of no value for every child, regardless of age, type, distribution of the condition, severity of the physical handicap, extent of associated sensory and perceptory involvement, and degree of mental impairment. No one would dispute that great difficulties are involved in attempting to compare the value of any kind of treatment with that of another. One might question, however, whether the difficulties are sufficient in themselves to justify neglecting to make some attempt at assessment, which at least should prove somewhat more constructive than the uncertainty which at present exists.

The definition of the term 'cerebral palsy' is 'brain damage acting on an immature brain'. This means that two factors have to be considered in the management of the child with cerebral palsy—

- (1) The effect of the lesion on the process of maturation—i.e., its influence on the orderly development of sensori-motor patterns in response to environmental stimulation and its effect on the physical and mental development of the child.
- (2) The pathological conditions produced by the lesion, including:
 - (a) abnormal qualities of muscle tone, such as spastic and plastic hypertonus, flaccidity or an unstable muscle tone;
 - (b) abnormal postural reflex activity;
 - (c) sensory and perceptory disturbances involving vision, hearing and proprioception, among others; and
 - (d) various degrees of mental subnormality.

The symptomatology of cerebral palsy is not stationary, and the diagnosis is often doubtful during the first few months. In a few cases the early signs of brain damage disappear and the children develop normally. However, such disappearance of symptoms usually occur not later than 18 months to 2 years of age, and more often before. In most cases symptoms develop gradually during the first three years, and may still change up to late adolescence. Spasticity and athetosis gradually increase, in spite of, and most probably as a result of, the maturation of the central nervous system.

In both the normal and the cerebral palsied child, sensory stimulation and experience in their widest application are needed for maturation. In the normal child they lead to normal development—that is, without further help (or treatment) the child responds to the sensory input with an appropriate and adequate motor output. He attains his milestones in the normal way and at the proper time by building up every new activity on the ones already

learned. He uses his existing 'learned' sensori-motor patterns in movement and speech, and changes and adapts them to his increasing and more selective functional activities. In this process of maturation the gradual acquisition of head control and equilibrium in all positions plays an important part.

With this in mind one might ask oneself: is it sufficient to rely on the process of maturation alone, by giving a young child with cerebral palsy the normal amount and normal quality of stimulation? Will this alone help him to develop to the utmost his potential sensori-motor patterns in a reasonably normal way? Can one expect reasonably normal motor responses from such a child without the help of physical therapy—i.e., without handling the child, guiding his motor output, and counteracting abnormal patterns of posture and movement, in order to give him the experience of the most important fundamental sequences of the normal postural control and movement which a normal child acquires during the first three years of life? Can the child learn to walk and use his hands in a fairly normal manner without developing contractures and deformities?

To some all this seems most unlikely because the cerebral palsied child cannot, if there are appreciable degrees of neurological disorder, react with normal motor responses to sensory stimulation. His maturation, however much encouraged by motivation, guidance and good home management, will be best be patchy and abnormal. If he is intelligent and able to move to some extent, he will acquire functional abilities but with abnormal motor patterns, which he will use as a basis for every newly learnt activity. Because of this the abnormal patterns will become reinforced by repetition, and he may in time develop contractures and deformities. Though it is true that no amount of treatment will make a child 'normal', this should not lead to the argument that as long as the child does what he is capable of doing the degree of abnormality does not matter, and can be dealt with later by bracing and surgery. If this were accepted, the conclusion might be drawn that treatment should only be given to the older child if he needs it to correct his faulty patterns of posture and movement and acquired contractures and deformities. Would it not be better to treat the very young baby, in whom spasticity or athetosis is still slight, and whose motor activity is still primitive rather than abnormal? By helping him to establish more normal sensori-motor patterns, one might allay the increase of spasticity or athetosis, and will certainly prevent the development of contractures and deformities in the majority of cases.

Many workers seem to think that children under 18 months of age, and older children if they are intellectually handicapped, cannot co-operate and make active movements in treatment. Therefore they rightly say that treatment by passive movement only will not give normal proprioceptive sensations such as active movements produce. This may be so when using the orthodox therapeutic techniques. We are often asked: How can you make babies move actively in any specific way? To this one can answer that special techniques of handling the child can stimulate and facilitate active motor responses to being moved—i.e., motor responses which a normal child develops naturally when being handled (washed, bathed, dressed, fed, picked up, put down, etc.) by his mother. In treatment, the child is handled in a way which makes him adjust his posture with spontaneous active movements, while at the same time abnormal motor responses are prevented. In this way he learns the most fundamental movement patterns of righting his head, adjusting the position of his limbs when his body is moved, keeping his balance, etc. He learns to co-operate when being handled instead of being passive. This gives him normal movement patterns which he can use later for voluntary movements.

Home management alone might benefit the child with a minimal physical handicap and

normal, or even subnormal, intelligence. In the more severely affected child it may prevent any further deterioration of the mental condition, but at the expense of neglecting the physical condition.

No type of physiotherapy will give satisfactory results without the co-operation of the parents at home. The treatment, especially if it does not consist of a number of set exercises but of normal functional movement patterns, has to be co-ordinated with, and incorporated into, the child's daily life, self-help and play. For this reason training and guidance of the parent is of the utmost importance, and this is best done if the mother or other members of the family are present when the child is being treated. It is also a good plan for the therapist to visit the child at home, at least once in a while, to see how he lives and what chances there are of applying the abilities he acquires in treatment to his daily activities at home.

The parents have to be taught to understand their child's difficulties and abilities at various stages of treatment and progress. The child has to have a wide range of experience, and if he cannot get around to explore his environment then it should as far as possible be brought to him. The parents should be helped towards a good emotional adjustment themselves, because without this the child will not be able to make his own adjustment.

Parent guidance and training by itself—that is, without treatment—is, however, inadequate except in those cases which show motor retardation with only minor neurological signs, or mainly mental retardation. The reasons for this are:

- (1) It needs the skill of a specially trained therapist to give the child a more normal muscle tone and the experience of new and more normal postures and movements, especially if his spasticity or athetosis is severe.
- (2) The mother should be trained to help the child to move in more normal ways once the patterns of these movements have been established in treatment and the child can use them with little help, but progress into new and more difficult activities will probably present problems which cannot be coped with by the parent alone.
- (3) The parents, who already have a great burden to carry, should not be loaded with the total responsibility for their child's progress or failure.

In conclusion, the problem is not really one of treatment against home management only, but treatment plus home management against home management only.

KAREL BOBATH, BERTA O. BOBATH

Minimal Cerebral Palsy

SIR—I was impressed by Dr. Wigglesworth's and Dr. Walton's letters in the June and August *Bulletins* (pp. 293 and 391). I am quite convinced that this condition of minimal cerebral palsy does exist and that it is important to call attention to it. I would like to add some of my personal experiences with so-called clumsy and backward children during 11 years in an outpatient clinic for neurological disorders in children. This experience supports and in some ways completes Dr Wigglesworth's views.

Delayed development of the central nervous system is one of the signs of cerebral palsy. In the minimal cases the condition may be limited to delayed development, and this may be a selective delay, involving motor and/or speech development only. But I believe the developmental retardation may be even more selective, as in cases of '*delayed development of extrapyramidal motility*'. These children sit, walk and talk at the proper times but are strikingly clumsy. All motor performance which requires involuntary movements is difficult. They are bad at games and fail wherever skilled movements are required. They

show no signs of spasticity and a very careful examination reveals only slight extra-pyramidal signs: asymmetry of postural tone, with some involuntary movements which increase with fatigue. But the past story reveals some difficulty in labour and some degree of asphyxia or prematurity. The situation of such a child is not always a happy one. His schoolteachers and family are rarely sympathetic, and the child may even be despised by his schoolmates because of his unfitness for games, so that a neurosis is added to his motor disability. On the other hand, early specialised exercise under the guidance of a child neurologist may completely restore the motor abilities of these children. Some of them can train themselves and make up for their developmental delay, but in any case it is necessary that not only paediatricians but also schoolteachers should understand that the clumsy child may be suffering from a form of 'minimal cerebral palsy'.

There is a second point: Dr. Walton referred to the apparently mentally retarded children in whom defects of an apraxic and agnosic character were found, but there is another reason for the backwardness of physically handicapped children. This is the lack of external stimulation—the lack of experience. This happens when the child is kept at home without regular physical exercise or medical treatment in a family with a large number of children. The child who does not move about cannot acquire sufficient experience and may seem to be mentally retarded. We have observed such children who, with a proper training and medical and physiotherapeutic care, have overtaken their backwardness in about 3 months and caught up with their chronological age.

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IVAN LESNÝ, M.D.

(*Editor's note: we shall be publishing an original article on Minimal Cerebral Palsy by Dr. Richmond Paine in our next number.*)

Speech Therapy for Developmental Disorders

SIR—I hesitate to prolong a correspondence which many of your readers must feel has gone on long enough but one point in Miss Renfrew's letter does require amplification.

It is true that I think that speech therapy is more often of value in the child of school age than in the pre-school child. At the same time I think it should be realised that a great deal of unnecessary, though probably harmless, speech therapy is being undertaken in our schools. As Dr Mary Sheridan has pointed out on a number of occasions, doctors seem peculiarly uninterested in speech disorders in childhood. The result is that children are referred direct from teachers to speech therapists in many cases and speech therapy is given without any adequate prior medical or psychological assessment.

All too often the speech therapist is only nominally attached to school medical or psychological services and in fact is left to diagnose and treat her patients almost single-handed. An experienced speech therapist like Miss Renfrew can clearly cope with these duties far better than any doctor not employed largely on examining patients with speech disorders but a great many inexperienced therapists are unable to distinguish patients who require treatment from those who do not. If adequate assessment of children with speech disorders was made more often a large number of children who at present receive speech therapy but do not need it would be removed from speech therapists' treatment lists. The speech therapist would then be in a better position to treat children who really do need speech therapy more intensively.—Yours, etc.

Department of Child Life and Health, University of Edinburgh

T. T. S. INGRAM

BOOK REVIEWS

Cerebral Palsy in Childhood and Adolescence

Edited by J. L. HENDERSON

Professor of Child Health, University of St. Andrews

Edinburgh and London: E. and S. Livingstone, 1961, pp. 385, 35s.

Reviewed by RICHMOND S. PAINE, of Boston, Mass., U.S.A.

This is one of the most thorough regional surveys of cerebral palsy made in recent years. It was planned to study all cerebral palsied patients under 21 years of age in the Eastern Hospital Region of Scotland, cases being found through Medical Officers of Health, hospital records (including search of possible related diagnoses), institutions for chronic disease, and finally contact by letter with all general practitioners in the area. All patients were to see the social worker and psychologist at least twice and to have one or more examinations by the paediatrician, orthopaedic surgeon, otolaryngologist, ophthalmologist, dentist, and blood-typing laboratory. Electroencephalograms were contemplated but abandoned. From a screening list of 323, 240 cases were confirmed; 22 had died or moved away, and 61 had other final diagnoses (36 of these being mental retardation alone, reflecting discernment in this respect). Three years of examining were required, and staff members travelled over 29,000 miles, but the survey was accomplished at a modest cost of £30 per confirmed case, doubtless because 11 of the 14 man team contributed their services on an 'honorary' basis. The data obtained are most impressive in view of the size of the budget, the dispersed population apart from the cities of Dundee and Perth, and the fact that 43 per cent of the mothers in the area are employed.

The definition of cerebral palsy adopted is an accepted one ('A disorder of motor function resulting from permanent non-progressive defect or lesion of an immature brain'), but no upper age limit was imposed as to onset. The classification of the American Academy for Cerebral Palsy was used, with some modifications, but only 5 of Phelps' 12 modalities of athetosis are retained, which will impress some as still too many.

Most of the conclusions add little new to other surveys: the prevalence-rate of about 2/1,000 in age range; the distribution by types (9.6 per cent as mixed agrees with recent surveys but not with many earlier ones); 27.5 per cent with birth-weights under 5½ lb.; 25 per cent with epilepsy (but more frequent in spastic diplegia than in hemiparesis, unlike some other reports); 48 per cent with I.Q. under 70; 25 per cent with behaviour problems; and 10.4 per cent in institutions. There are, however, several areas of divergence or of special interest: 60 of 153 weighing over 5½ lb. at birth had abnormal deliveries, with use of forceps in 25 per cent (compared with 6.5 per cent in random deliveries in neighbouring Aberdeen), and breech extractions in 4.6 per cent (*cf.* 1.2 per cent in Aberdeen). Surprisingly, no athetotics were breech births; 33 per cent had a history of abnormality early in their mothers' pregnancies, but this was three times commoner

if the birth was normal and at full term; and 12 per cent of all cases were considered to be postnatally acquired (50 per cent of the athetotic group: a high figure, possibly attributable to the small number—19—of athetotics with documented aetiological histories).

Inquiry was made as to the occurrence of dystonic spasms in the evolution of the clinical picture; the incidence obtained (5.8 per cent) would inevitably be on the low side of fact, and the authors correctly state that the spasms are not confined to any one type of cerebral palsy.

Another interesting figure is the 58 per cent incidence of ophthalmological abnormalities, an area in which this is one of the most thorough of all surveys. The 20 per cent found to have hearing losses, is comparable with Fisch's figure, but far above the 5-7 per cent of most other surveys.

Of the cases over school-leaving age, 53 per cent were thought potentially

employable, but significantly, only 60 per cent of these were actually employed.

The final chapter on recommendations reflects thoughtful and intelligent utilisation of the results of the survey for future planning of treatment, education, and care in the community, although some readers will question the emphasis on intensive therapy of the mentally deficient. It is surprising, too, to read earlier in the book that 'tendon lengthening is of doubtful value in spastic paralysis' and that 'only one of 37 such operations gave a satisfactory result'.

The book is understandably replete with statistical tables, but less understandably encumbered with individual case-histories. Yet it makes interesting and lively reading. It merits a high place among the several recent surveys of cerebral palsy in communities, and is recommended to those working or interested in the field, whether physicians, psychologists, educators, therapists, or social workers. R. S. PAINE

Molecular Genetics and Human Disease

Edited by Lytt I. Gardner

Springfield, Ill.: Charles C. Thomas, 1961, pp. 297, 92s.

During the past 50 years astonishing progress has been achieved by man in his ability to understand the properties of inorganic matter. We seem now to be on the threshold of a similar revolution in the biological sciences, whereby biological processes, including the fundamental one of reproduction, are being comprehended in terms of molecular structure.

The biochemists have at last begun to achieve success in what had seemed the hopeless task of elucidating the detailed chemical structure of the proteins, including the crucially important DNA, the raw material of chromosomes. At the same time, by the simple inspection of tissue cells under the microscope (an approach which until recently had seemed unlikely

to expose fundamentally new facts) individual chromosomes can be seen, counted and mapped, so that various clinical syndromes are now recognised to be the result of identifiable chromosome patterns.

Recent progress in genetics, with particular reference to the molecular structure of proteins, and to the chromosome basis of disease, was discussed by 14 eminent scientists, about half of them medical, at a symposium held at the State University of New York at Syracuse in April 1959. All except two contributors, namely F. H. C. Crick and Paul E. Polani from this country, came from North America. The meetings were evidently informal and the discussions, which are reported verbatim, lively and to the point, a quality

which does not necessarily always result from such a bringing together of workers from different disciplines.

This book is essentially one by specialists for other specialists in this field. It is well

printed and produced, and the only criticism which could be levelled at it is the fact that over a year and a half has elapsed between the symposium and this account of it. DOUGLAS GAIRDNER

Chemical and Molecular Basis of Nerve Activity

By DAVID NACHMANSOHN

New York and London: Academic Press, 1959, pp. 235, \$7.50

The recent introduction of the word neurochemistry to our scientific vocabulary gives a measure of the interest now being taken, and the advances being made, in this branch of biochemistry. This monograph, by an acknowledged leader in this field, gives a useful and well-written summary of the work leading to present conceptions concerning the part played by acetylcholine in nervous processes. The author puts forward his view that 'the action of acetylcholine is not an *inter-* but an *intra-*cellular, or rather an intramembranous process essential for the generation of bioelectric potentials in all conducting membranes throughout the animal king-

dom'. While we may not all be in complete agreement with some of the interpretations that he has put on his findings, and although some aspects of the field are dealt with narrowly, this is a book which is unquestionably stimulating, and should certainly be read by all who are interested in the problems presented by nervous conduction and transmission.

The chapters dealing with acetylcholinesterase and with its inhibition by organo-phosphorus compounds, together with the work leading up to the introduction of compounds capable of reactivating the inhibited enzyme, are full of useful information. R. H. S. T.

Tumours of Childhood

By H. W. DARGEON

London: Henry Kimpton, 1960, pp. 476, £7 10s.

This book is divided into two parts. The first is entitled General Considerations, and in it such subjects as incidence, diagnosis and treatment are discussed. Discussing the early recognition of cancer, the author is of the opinion that frequent medical examination of children is essential. He even states (page 43): 'during infancy rectal examination should be performed at three months, or earlier if indicated. Subsequently, a routine rectal examination at six monthly intervals up to the age of four years would detect many of the pelvic tumours'. This advice is fortunately not likely to be followed. Considerable space is devoted to radiotherapy, and there is a

valuable section on the chemotherapy of malignant disease. Psychological management is also discussed with sympathy.

The second part of the book is on *Specific Tumours*. The word tumour has been interpreted with surprising liberality to include all new growths, malignant and benign. For instance, leukaemia, simple naevi, lipoidosis and polycystic kidney are all discussed. Throughout, the emphasis is clinical and medical. Pathology and surgery are given little space. Of particular interest to *Bulletin* readers are the chapters on *Tumours of the Central and Peripheral Nervous Systems*. Thus in the section on von Recklinghausen's disease, the protean

nature of the condition is stressed and the importance of considering it in the differential diagnosis of any progressive nervous disorder is emphasized. It is stated that café-au-lait spots are present in 90 per cent of cases of neurofibromatosis, but in only 10 per cent of normal individuals.

The book is profusely and gruesomely

illustrated and well written. The wideness of the subject-matter means that no topic is really adequately covered, but this is compensated to a certain extent by an extensive bibliography. This is a useful reference book, particularly for those requiring advice on non-operative treatment of cancer in children.

JOHN RENDLE-SHORT

An Introduction to Human Blood Groups

By FULTON ROBERTS

London: William Heinemann, 1960, pp. 182, 9s. 6d.

Knowledge of blood-groups tends to be fragmentary. Paediatricians manage erythroblastosis irreproachably but are hazy about genetics; geneticists conjure with complexes but know little of kernicterus; physicians argue about associations; anthropologists sail their own course and immunologists are increasingly erudite. Dr. Fulton Roberts in 85 pages effects a synthesis so that all may profit.

In Chapter 1 he introduces us to serological terms and in five pages tells us the basic principles of antibody formation. He then devotes a third of the book to the Rhesus blood-groups and it is here that he is at his best for the simple reason that he takes us slowly. A point in this system which is not generally appreciated is that D is not 'dominant' to d but only a much more powerful antigen. In the actual syndrome of erythroblastosis the twin dangers are anaemia and kernicterus—these to some extent independent of each other in that immaturity of the liver in conjugating bilirubin can be as much a factor in the production of jaundice as a high antibody titre; furthermore, iatrogenic haemolysis from vitamin K overdosage must not be forgotten.

Complete and incomplete antibodies are next explained, and anyone who cannot understand and remember the Coombs' test after his lucid exposition had better

give up medicine. In the genetics of Rhesus, crossing-over is explained and one then sees a possible reason for the three degrees of commonness of the various genotypes.

Dr. Fulton Roberts deals with the ABO groups almost last because he feels they are not typical and not simple. Thus naturally occurring antibodies are present, the antigens are often found in body fluids as well as on the red cells and suppressor genes may occur. In an interesting way he contrasts the difference between the immunisation of a mother by an incompatible Rh foetus and by one incompatible on the ABO system. In Rh incompatibility, transplacental passage of foetal cells is necessary, the antibodies are 'incomplete' and the Coombs' test is positive. In ABO incompatibility, water-soluble antigens, not cells, pass through the placenta and it is these which invoke immune antibodies; returning to the foetal circulation they are responsible for the haemolysis in the infant, but for some ill-understood reason the Coombs' test is negative.

The author is most emphatic about the importance of cross-matching in routine blood-transfusion. Wherever blood for transfusion is itself available there must be facilities for accurate grouping, and when in doubt or in a hurry, giving O dd blood is *not* the answer. This is because:

1. There are limits to what the blood-transfusion service can supply.

2. Group-O blood sometimes contains anti-A or anti-B in such high titre that its effect is damaging to the patient's cells, particularly if the antibodies are of the immune type. Such blood he thinks should be called DUD (Dangerous Universal Donor).

Some other blood-groups besides Rh and ABO are briefly mentioned (the Lewis system in less than three pages would be somewhat troublesome for a beginner), there are three pages on the association of the ABO groups with disorders of the gastrointestinal tract, and there is a chapter on blood-groups and anthropology

which would be all the better for a map.

The author concludes with some 'growing points', and he here discusses the chemical constitution, distribution and affinities of antigens and antibodies. Antigens are to be found increasingly on cells other than red corpuscles, while antibodies are related to other inherited gamma-globulins, of which the haptoglobins and Gm groups are the best known examples.

The only adverse criticism of this comprehensive and excellent book is that occasionally in some of the shorter chapters one wishes the subject-matter were not so condensed.

C. A. CLARKE

Multiple Sclerosis: Prognosis and Treatment *A Nosometric Approach*

By LEO ALEXANDER, AUSTIN W. BERKELEY AND ALENE M. ALEXANDER
Springfield, Illinois: Charles C. Thomas, 1961, pp. 188, 60s.

Any information on the natural history of this condition must be of value, and in particular, any attempt to overcome the notorious difficulties in assessing the effect of treatment. The authors of this monograph use a numerical method, and, although this is still open to the possibility of subjective error, the risk is obviously minimised by covering as many aspects of the disease as possible. The careful analysis of several hundred patients followed over a number of years helps to confirm many clinical impressions. For instance, the greater the degree of recovery from a preceding attack, the more likely are subsequent ones to have a favourable prognosis. A steady deterioration in the patient's condition is likely to lead to the gravest disability and this has led the authors to postulate a recovery or resistance factor which may differ markedly from person to person. Evidence in favour of this included the good prognosis that was found among patients who had a severe

attack in the early stages of the disease if this was followed by prompt recovery.

It has been considered in the past that blood-transfusion from a healthy donor might restore some missing factor and this line of treatment was therefore tried. It was found that the patients most likely to benefit were those who had attacks which did not start to recover within a reasonable time or terminated after only a partial improvement.

Adrenocorticotrophic hormone was used with apparent success when the disease was developing fairly rapidly with the occurrence of frequent attacks and when the progression of the disease due to overgrowth of scar tissue was tending to continue during the intervals between the attacks. Cortisone and Prednisone did not prove affective and it was suggested that this might indicate that the adrenal hormones responsible for improvement under the stimulating effect of ACTH might be hormones in addition to cortisone.

Seventy-six patients were treated with transfusion and forty-three with ACTH and careful controls were used. However, although in both cases the course of the disease appeared to be favourably affected, it would seem that in the event of improvement rather than cure these claims will

have to be checked by a number of different workers before they can be fully substantiated. Nevertheless, this is an interesting and stimulating book on a subject which is so often depressing for those who come into intimate contact with it.

NEIL GORDON

The Eye in General Practice

By C. R. S. JACKSON

Edinburgh and London: E. & S. Livingstone, 1960, pp. 152, 21s.

Until the appearance of the first edition of this book, there was no good, brief guide to ophthalmology, and this simple textbook for undergraduate students and G.P.'s has filled a wide and deserved market. In spite of a discursive style, there are not many omissions, although one looked in vain for word of some major conditions like nystagmus. The only serious criticism concerns the illustrations. They are far too few, so that most of the descriptions (as of central artery occlusion, acute glaucoma, anatomy of the lacrimal passages, conjunctival follicles and phlyctens) cry out for a picture in vain; and others, like acute iritis, are dismissed with one indefinite picture (that shows no K,P. and no obvious festooning or ciliary

injection—in fact, only a small pinkish eye with a large pupil, surrounded by a lot of pink face). The advantage of having the majority of the illustrations in colour is entirely outweighed when these are of such terrible quality. The dendritic ulcer, for instance, shows as a vague black dendritic line with a vague green surround (!) against a violet iris and purple pupil. If only these were mostly in monochrome they could be clearer and far more numerous, and the book would be a little less expensive. We can only hope that the publishers will redeem their good name when the time comes for the third edition of a book whose place is clearly established in the medical curriculum.

P. D. TREVOR-ROPER

Pediatric Pathology

By DANIEL STOWENS

London: Bailliere, Tindal & Cox, 1959. Baltimore: Williams & Wilkins, 1959, pp. 676, £8.

The field of paediatric pathology is so wide and involves so many special branches of pathology that to produce a textbook to cover the whole field adequately is perhaps impossible. That there has been no real attempt to do so until this book is itself a measure of the difficulties and the energy and enterprise of Dr. Stowens. The result is a beautifully produced volume with a large number of fine photographs, an excellent clear typography, and an extensive bibliography and index.

The material used in the book appears to be derived almost entirely from that going through the Armed Forces Institute of Pathology in Washington. This has ensured the advantage of the inclusion of excellent photographs from many rare conditions but at the same time a relative lack of touch with the day-to-day problems of pathology in childhood.

The approach of the book is encyclopaedic and theoretical. Thus reference may be found in the book to almost all the

rarer diseases of children, but the comments on these are often no more than can be found in a purely clinical book.

The section on nervous diseases is one of the best, but there is only one page on infantile cerebral palsy. In this the author stresses the many types of underlying pathology and also the gaps in our knowledge of the structure of the newborn brain. The section on the brain includes a number of full-page microphotographs, the point of which is sometimes difficult to see. Other sections of the book are highly variable, those on diseases caused by protozoa and spirochaetes being good,

while the section on sudden and unexpected death is very narrow in outlook.

Dr. Stowens is to be congratulated on producing the first modern comprehensive book on paediatric pathology for the general pathologist, who will find it chiefly useful as a source of references to relatively rare diseases. The paediatrician will find little more in this book, other than good microphotographs, than is available in special clinical monographs. The paediatric pathologist will be more than ever convinced of the difficulties of writing a good general textbook on the pathology of paediatrics.

J. EMERY

A Guide for Child-Care Workers

By MORRIS FRITZ MAYER

New York: The Child Welfare League of America, 1958, pp. 184, \$2.25.

The Child Welfare League of America assigned Dr. Morris Fritz Mayer, because of his vast experience and understanding in the field of child-care, particularly in institutions, the task of writing this study of the child's normal emotional development, and of the problems that will arise for the child living away from his parents in institutional care. This book will give invaluable help to all concerned in the day-to-day care of children, and it will give an insight into the way integrated efforts can be made to enable the child to establish relationships with those around him, to achieve parental identification, and to develop his full potentialities so that he can be happy in the everyday life of Society.

An excellent chapter gives a concise account of the basic needs of all children for their healthy physical, emotional, social and intellectual development and deals with the particular needs of children in institutions. The cause of the various problems which arise in different age-groups, and their management, is discussed.

The emotional interaction of the child-care worker and the children with their different personalities causes strains which must be appropriately handled so that group living may be balanced and enriched.

We are taken through the day from getting up to going to bed. No matter on what social or philosophical beliefs institutions are based, all who run them must encounter similar difficulties. Advice is given as to the way in which these difficulties arise—thus the significance of illness, the part age plays in the standard of cleanliness and clothing and the allocating of household chores are all considered. It is the supreme assignment of the worker to become the home-maker. Every opportunity must be taken to build an atmosphere of affection and understanding upon which all good relations are made and once in this situation the child will become more amenable to discipline. Punishment given must be logically understood to be effective.

The contributors to this book are alive to the necessity of all within the team making

their contribution to child care with co-ordination. Workers in the field must be given greater opportunities to receive training for this is a field where the

experience of others can help to enrich one's own experience.

M. THEISEN, S.R.N., D.N. (Lond.)

Health Visitor

The Royal Edinburgh Hospital for Sick Children 1860-1960

By DOUGLAS GUTHRIE

London and Edinburgh: Livingstone, 1960, pp. 75, 17s. 6d.

This is the story of the founding and first 100 years' work of one of our most famous children's hospitals. As we would expect of its author, who incidentally is not a paediatrician, the book is delightfully written. It is strong ammunition for those who are resisting the present trend in some high places to play down the need for special children's hospitals. In fact, the idea of founding such a hospital in Edinburgh 100 years ago did not meet with universal approval. Dr. Guthrie, indeed, makes the point in the first page that 'such erroneous views not only delayed the establishment of children's hospitals, but also retarded for many years the emergence of paediatrics as a special and separate branch of medicine'. Paediatricians are even now fighting a long campaign, aimed at convincing not only administrators but some of their own medical colleagues that children are not simply to be regarded as miniature adults. It is only in fully equipped

children's hospitals that the diseases and problems of infancy and childhood are likely to receive the special consideration which leads to real advances in knowledge and practice.

The pages of this book bear eloquent testimony to this view. Many famous names in medicine and surgery appear here—John Thomson, Charles McNeil, Joseph Bell, Sir Harold Stiles, and Sir John Fraser were all members of the staff. Prof. R. W. B. Ellis, the present holder of the University Chair, contributes a chapter which focuses attention on the problems facing all children's hospitals today, and makes a plea for recognition of their essential part in research, teaching and service to the community.

This book should be read by all who have any interest in or responsibility for the hospital services of this country in the present era of rapid change.

J. H. HUTCHISON

The Practice of Clinical Child Psychology

By ALAN O. ROSS

New York and London: Grune and Stratton, 1960, pp. 275, 41s.

The psychological assessment of the child may still be more of an art than a science, yet even in art there are certain rules of procedure, do's and don'ts, the universal application of which are of help in gaining understanding and in sharing of knowledge.

In this book an attempt has been made to spell out these rules in detail, and its primary virtue lies in the balance which

the author has been able to maintain throughout between the scientific and the intuitive aspects of the psychologist's work. Thus a detailed examination of psychometric tests is interspersed with warnings about the danger of inflexibility in applying such tests; stress on using tools of proved reliability and validity is followed by the warning not to interpret quantitative test results too literally; and the need for more

research on projective techniques appears side by side with a plea not to be paralysed into clinical inactivity until the research material has all been gathered. From this point of view the book makes an excellent teaching manual, and it is indeed mainly to students of clinical psychology that the author has addressed himself. Other readers are likely to find the book rather too detailed to capture interest, and the rather uninspired level of writing and the long, self-conscious and axe-grinding discussion of professional role and relationships with other professions are further handicaps in this respect. It is also a pity that no material on the assessment of the very young child has been included: many psychologists working in just the kind of setting described by the author will be surprised at his assertion that the under-three year group is rarely seen by them. A rather egocentric trend is revealed thereby, which is further accentuated by the description of clinic practices. These tend

to differ far more from clinic to clinic than the reader will be led to believe by the present account, and a rather wider frame of reference is clearly indicated.

Considerable discussion, including two detailed case studies, is given to the problem of evaluating children with cerebral pathology. A theoretical framework for such evaluation is presented, but as this is entirely based on the single concept of *integration*, it is unlikely to attract much interest. The description of the way in which psychological tests can elucidate cerebral pathology is, however, excellent and represents probably the best part of the book. The interaction of emotional and organic factors, the relation of psychological to neurological findings, and the details of the contribution of different tests to diagnosis are all presented in a very competent fashion and make this section well worth reading.

H. R. SCHAFFER

The Education of Slow Learning Children

By A. E. TANSLEY AND R. GULLIFORD

London: Routledge & Kegan Paul, 1960, pp. 235, 28s.

This book is explicitly intended as a text for the teacher specialising in backward children, and the medically qualified reviewer therefore approaches his task with a certain diffidence. Like many books, it reveals as much about the authors as their subject. The worker in related fields who has not already discovered how high an ethical standard the teaching profession sets for her better-paid sisters will be impressed with the practical idealism implicit in their approach. This could be summed up in the Marxist slogan 'to each according to his needs, from each according to his ability'.

The opening chapters would be profitable reading for anyone—be he parent, administrator or doctor—concerned with

apparently backward children, but thereafter the book becomes repetitive, the authors tending to make the same points under different headings as if their readers as well as their subject were slow learners. The chapter on physical disease as a cause of backwardness, while a fair summary of established knowledge, misses the opportunity of interesting the teacher in present trends of medical and psychological research: it presents the interested physician with an obvious challenge. In the sections dealing with preparation for adult life the handling of sexual adjustment might have been expanded and the approach is a little too moralistic: if these children have difficulty with conceptual thinking and interpersonal relationships, they are likely

to find it even harder than the rest of us to appreciate the need for postponing, sublimating or cannulating their instinctive drives. One clear lesson emerges: that the association between backwardness and emotional disturbance is due to maladaptation rather than to both being a result of brain damage or inferiority, and that in a suitable environment a good proportion of

these children can grow into responsible adults despite intellectual limitations. As regards the book's value to the intending teacher, it seems to lack detailed instruction in technique, but there is an excellent bibliography and it would be an ideal introduction to the subject. It is well produced and this reviewer did not notice any important errors. J. A. DAVIS

Emergency Syndromes in Pediatric Practice

By ALFRED J. VIGNEC

New York: Landsberger Medical Books Inc., 1959, pp. 382, 70s.

An ambitious attempt is made in this book to describe illness in childhood in which there is no time to think, no time to indulge in diagnostic aids and no time to allow the picture to clarify itself; this includes much more than acute emergencies and indeed involves much of general paediatrics.

In the first section, management of respiratory distress of the newborn is described with an excellent account of the quick but detailed assessment which is so important. Treatment is much more active than is the rule in most British centres; for example, when the infant is white and limp from the beginning, endotracheal intermittent positive pressure is regarded as a routine procedure. Most of us would agree with the author's suggestion, in his account of respiratory difficulties in older children, that the term croup should be abolished. In acute asthma treatment with long-acting adrenaline and with rectal and intravenous aminophylline is recommended, and in addition A.C.T.H. gel for a period of five days or in some cases oral steroids. Most British paediatricians try to avoid using steroids in asthma because once started they may be difficult to stop.

There is an excellent account of the management of cardiac failure in the section on cardiovascular diseases and there is also an account of routine investiga-

tions which cannot really be regarded as 'emergency'.

Dr. Vignec's chapter on blood diseases is a clear exposition of a subject which tends to be confused by variations in terminology; there is a particularly good account of exchange transfusion.

The section on gastrointestinal disease is divided into four sections, all of them most valuable. Emergencies in the newborn period, particularly obstructions, are followed by an account of acute conditions in the first two years and then in later childhood. The importance of appendicitis is emphasised and Gross' figure of 80 per cent of cases of appendicitis having perforated before operation in children under four is quoted to underline the importance of acute abdominal emergencies in this age group.

The section on the central nervous system contains a good account of the emergency treatment of convulsions; the author prefers to use sodium phenobarbital 1 grain per year of age up to five years as emergency treatment, with as an alternative rectal Seconal (quinalbarbitone), using capsules rather than suppositories. Paraldehyde, which is much used in this country, is not mentioned. On the possibility of meningitis and the question of lumbar puncture, he says: 'Unless there are severe compelling contraindications

this examination should always be made.' Most of us would agree that unless lumbar puncture is the rule early cases of meningitis will inevitably be missed.

The chapter on metabolic disorders is perhaps the most valuable in the book, particularly the detailed reference tables of fluid and electrolyte requirements; pleasant too to find a modern book which does not assume that everyone now thinks in terms of milliequivalents. The initial dosage of insulin in diabetic coma in childhood is described as one unit per pound of body-weight. The child, however, is commonly brought in in pre-coma and it is most important to remember that in these cases a small initial dose only should be needed.

There is a good account of burns, which are included under metabolic disorders. The separate chapter on steroid therapy is useful in summarising the present position and is followed by one on poisons, which will be much referred to. The book ends with chapters on premature infants and—inevitably in an American book—one on allergy.

This book is written in a simple and direct manner with great economy, so that a large amount of material has been included in its 370 pages. It will be a valuable work of reference in the paediatric department, where the sort of information it contains is often both urgently needed and difficult to find. J. J. KEMPTON

In Memoriam

XLV

The baby new to earth and sky,
What time his tender palm is prest
Against the circle of the breast,
Has never thought that 'this is I:'

But as he grows he gathers much,
And learns the use of 'I' and 'me',
And finds 'I am not what I see,
And other than the things I touch.'

So rounds he to a separate mind
From whence clear memory may begin,
As thro' the frame that binds him in
His isolation grows defined.

This use may lie in blood and breath,
Which else were fruitless of their due,
Had man to learn himself anew
Beyond the second birth of Death.

Alfred, Lord Tennyson.

ABSTRACTS

In collaboration with *Abstracts of World Medicine*, published by the British Medical Association, and with the kind assistance of the Excerpta Medica Foundation, *Courrier*, and *Obstretical and Gynecological Survey*.

The Dental Problems of Handicapped Children

DON CHALMERS LYONS. *Children, Washington, D.C.*, 1961, 8, 14-16.

Dr. Lyons has considerable experience in the dental care of handicapped children, especially those suffering from cerebral palsy. In this article he stresses the social importance of the teeth, particularly to the handicapped. He points out that in the United States dental treatment for these children is an almost unsolved problem. What is done is often little more than the emergency relief of pain by indiscriminate extraction of teeth. This aggravates the dental conditions present and adds to the problems that they produce for the patient. Dr. Lyons feels that lack of good oral hygiene undoubtedly contributes to the high incidence of dental caries (decay) in many handicapped children, especially those with cerebral palsy. The parents may try to do a good job, but they need special training, and the children need assistance, perhaps in the form of a mechanical toothbrush. The children's diet, too, needs attention, for although it may fulfil all nutrient requirements, if the food is soft, semi-liquid and sticky, particles pack round the teeth and cause decay. This type of food is given because it is easiest for both child and parent to deal with, but if training clinics gave more attention to helping the children to use specially constructed utensils and master techniques of chewing, they might manage a more

fibrous self-cleansing dentally acceptable diet.

Summing up the role of the dental services in the prevention and treatment of dental diseases, Dr. Lyons points out that few are willing or able to carry out, on a purely altruistic basis, the continuous programme needed for these children. Adequate dental care would cost about \$190 (£63) a year per child. The lack of facilities for this type of work is due in part to the shortage of dentists in general, a shortage which if unchecked would result in there being in 1975 only 50 dentists per 100,000 persons in the U.S.A., compared with 57 in 1958. (In Great Britain there about 50 dentists per 156,000 persons.) To meet this need more dental schools are to be built in America, and efforts are also being made to increase the ability of dentists to deal with handicapped children by postgraduate training.

In conclusion, Dr. Lyons says: 'The majority of these handicapped children can be trained to have a productive life by being taught to use what they have. Any complete rehabilitation programme must include consideration of dental welfare. For a child to have to learn to live with his handicaps is enough of an emotional stress without having the additional burden of preventable and correctable disfiguring dental destruction, crooked teeth, pain and chronic debilitating infection from infected teeth.'

J. N. Swallow

Mongolism Related to Emotional Shock in Early Pregnancy (In English)

D. H. STOTT. *Vita humana* (Basel), 1961, 4, 57-76, 29 refs.

At the University of Glasgow the relationship between emotional shock in early pregnancy and mongolism was studied in the replies to a comprehensive questionnaire sent to the parents of 739 mongol children and 400 non-mongol retarded children. The questions related particularly to shock or injury to the mother during pregnancy and the type of such shock.

The incidence of emotional shock during the second and third months of pregnancy leading to a mongol birth was nearly twice as high as in any other month. The preponderance of shock during the first trimester was highly significant when compared with the incidence for the non-mongol births and with the theoretically even chance of shock at any time. In pregnancies leading to non-mongol defective children the incidence of emotional shock was highest during the second trimester. The incidence of physical injury was highest during the second trimester both for mongols and non-mongols.

The tendency to increased incidence of noxious agents during the later non-critical months—observed also in studies of rubella and influenza—and the possibility of a miscalculation of the date of conception are discussed. The concentration of psychogenic shocks during the second and third months of pregnancies leading to mongol births was noticed particularly when the shocks were 'lasting' rather than when they were 'ephemeral'. Suggestions are made for correlating these findings with recent work on chromosome anomalies in mongols and other defectives.

E. H. Johnson

A Longitudinal Study of the Growth and Development of Prematurely and Maturely Born Children. Part VII: Mental Development 2-5 Years

C. M. DRILLIEN. *Archives of Disease in Childhood*, June, 1961, 36, 233-240, 11 refs.

This paper from the University of Edinburgh continues a study of the growth and development of prematurely born children. Of the original 600 children (premature, with full-term controls) 92 per cent of the survivors remained in the survey at 3 years and 90 per cent at 4 years. They were examined at 6-monthly intervals up to the age of 2 years and annually thereafter. Mental tests were those of Gesell and of Terman and Merrill. About half the group have now entered school.

As at 2 years, mean scores on intelligence testing fell steadily at 3, 4, and 5 years with increasing birth weight. Twins had consistently lower scores than singletons of comparable birth weight. At 4 and 5 years there was a striking excess of children with a birth weight under 4½ lb. (2 kg.) who were unsuitable for education at school or were educationally subnormal.

The author states that the expected response to pre-school developmental tests is affected by birth weight, environment, and opportunity. She found (as others have done) that it is much easier to predict mental dullness in infancy than mental superiority. Of 16 children considered to be unsuitable for education at an ordinary school (I.Q. less than 70) 12 were assessed at this level at 6 months and at every subsequent examination; no child scored higher than 'very dull' at any test at any age. Of 16 considered 'border-line' defective in school (I.Q. 70 to 79) 12 were rated at this level or lower from the age of 6 months.

R. S. Illingworth

Effect of Bilirubin Metabolism in the New-born of Sulfisoxazole Administered to the Mother

H. I. KANTOR, D. A. SUTHERLAND, J. T. LEONARD, J. H. KAKHOLZ, N. D. FRY, and W. L. WHITE. *Obstetrics and Gynecology*, April, 1961, 17, 494-500, 35 refs.

The authors of this paper from St. Paul's Hospital, Dallas, Texas, had previously found sulphafurazole (sulfisoxazole, 'Gantrisin') a useful drug in the prevention of amniotic infection when labour was delayed following rupture of the membranes. Disturbed by the recent evidence that sulphonamides administered to newborn premature infants may increase the risk of kernicterus, they investigated the effect of maternally administered sulphafurazole on the bilirubin metabolism of the newborn child, and present the results of estimations of free and conjugated serum bilirubin in 278 normal infants, 44 of whose mothers had been treated with sulphafurazole. The dosage of the drug was: 4 g. initially, 2 g. 4 hours later, and then 1 g. 6-hourly until delivery.

In the 234 infants whose mothers had been untreated, free and conjugated bilirubin levels were estimated on cord blood and, in about a quarter of them, on specimens taken at one and 2 days old. Statistical analysis indicated that there were no significant differences between the results in those infants who had the same ABO or Rh (D) groups as their mothers and in those whose blood groups were different.

In the 44 infants of treated mothers, in which group most of the readings were made at one and 2 days as well as on cord blood, the conjugated bilirubin levels did not differ significantly from those of the controls, but the free bilirubin levels were distinctly lower. The maternal bilirubin levels were unchanged during the course of the treatment. Cord-blood levels of free

sulphafurazole were about two-thirds of the maternal levels at delivery, but elimination of the drug was almost complete after 2 days.

The authors suggest that the lower free bilirubin in the 'treated' cases was due to preferential binding of sulphonamide to serum albumin and a consequent transfer of bilirubin to the tissues—a situation which could be dangerous if the free bilirubin levels were abnormally high. They briefly mention current views on the metabolism of bilirubin in the newborn infant, and emphasise that the level of free bilirubin in the serum may not reflect the tissue level, owing to variations in the bilirubin-binding capacity of the blood. They suggest that in normal infants there is no evidence that maternally administered sulphafurazole adversely affects the child, but that sulphonamides or other drugs causing albumin-bilirubin dissociation should not be given if there is reason to anticipate that the child might be abnormally jaundiced after birth. *E. G. Hall*

'Uncomplicated' Hyperbilirubinemia of Prematurity. The Lack of Association with Neurologic Deficit at 3 Years of Age

J. G. SHILLER and W. A. SILVERMAN. *American Journal of Diseases of Children*, May, 1961, 101, 587-592, 17 refs.

In the premature nursery of the Babies Hospital, New York, 188 infants were examined clinically and the serum bilirubin level was determined daily during the first week of life. Of these, 110 were followed up for 3 years; of the remaining 78, 46 died (one from kernicterus) and 32 survived but were lost to follow-up.

The neurological and mental status of the children seen at 3 years of age was assessed, and a comparison made between those in whom in the neonatal period the serum bilirubin level was above 18 mg. per 100 ml. and those in whom the bilirubin

level did not exceed this value. There appeared to be no difference between the 'high' and 'normal' bilirubin groups in respect of the incidence of hypotonia, spasticity, speech defect, mental retardation, or behaviour disorders. In none of the patients were the classic features of kernicterus found. The authors suggest that premature infants may be able to tolerate high serum concentrations of bilirubin when this state is not complicated by erythroblastosis, administration of excessive doses of vitamin K, or administration of 'Sulfisoxazole' (sulphafurazole).

R. M. Todd

Measles Encephalitis. Evaluation of Treatment with Adrenocorticotropin and Adrenal Corticosteroids

S. KARELITZ and M. EISENBERG. *Pediatrics*, May, 1961, 27, 811-818, 14 refs.

The authors describe the course and treatment of 42 patients, 23 male and 19 female, admitted with measles encephalitis to the Queens General Hospital and the Long Island Jewish Hospital, New York, between 1952 and 1959, of whom 20 were followed up 2 to 7 years after discharge. The patients' ages ranged from 1 to 35 years, but the majority (30) were aged between 5 and 9 years. All but 3 were admitted within 24 hours of the onset of cerebral symptoms, at which time 23 patients were comatose, 15 lethargic, and 30 were having convulsions. In 41 cases the encephalitis became manifest 1 to 7 days after the measles rash appeared, in the majority (27 cases) on the 3rd, 4th, or 5th day; no case occurred in the incubation or pre-eruptive period. The time of maximum prevalence of measles was between March and June in the New York area.

Examination of the C.S.F. revealed wide variations. Thus the cell count ranged from below 20 to 650 per c.mm., lymphocytes predominating (numbers not given),

the protein content ranged between 50 and 197 mg. per 100 ml., and the sugar content from 48 to 197 mg. per 100 ml. The leucocyte count in the peripheral blood also varied widely, ranging from less than 10,000 to over 20,000 per c.mm., while differential count showed polymorphonuclear cells in the range of 40 to 93 per cent. The electroencephalogram was recorded on the 4th to the 30th day of illness in 19 cases and revealed abnormalities in 17. No attempt was made to isolate measles virus from the C.S.F.

Treatment was as follows. In 32 cases penicillin was given daily and 6 of these received chloramphenicol and 5 tetracycline in addition (does not given), while in 28 cases the patient was placed in an oxygen tent. Intravenous fluids were given for the correction of electrolyte imbalance. Tracheotomy was performed in 2 cases because of excessive secretions, and 3 patients received gamma globulin (12 to 48 ml.). Adrenocortical extract (4 to 12 ml.) was given to 3 patients, 6 received daily doses of ACTH (from 40 units diminishing to 5 units) and cortisone (300 mg. gradually reduced to 75 mg.), while 'several others' received the corticosteroids. Convulsions were treated by intravenous administration of barbiturates. All the patients survived. Emergence from coma or lethargy and cessation of convulsions occurred in most cases 24 to 72 hours after admission. Of 13 patients discharged with sequelae, 9 of whom had received steroids or ACTH, 3 developed behaviour defects, 3 mental deterioration, one schizophrenia and epilepsy, one muscular spasticity, and one paresis of the extremities. Of the 4 not given steroids, 3 manifested behaviour disturbances and one mental deterioration. Follow-up examinations carried out up to 7 years later showed that of 14 patients treated with ACTH and/or steroids, only 2 were completely well, 12 showing sequelae (7 emotional disturbance, 3

epilepsy, and one paresis of the lower extremities, which, however, was less severe than on discharge 2 years earlier).

The authors conclude that the use of ACTH and cortisone did not prevent neurological complications and possibly exaggerated them when present. They therefore do not advise these agents in the treatment of measles encephalitis and suggest that the prompt institution of the supportive regimen described above was the chief factor in the survival of all 42 of their patients.

(This paper is not clearly presented, but is nevertheless valuable in that it once again

demonstrates the dangers as well as the uselessness of steroids in viral infections.)

I. M. Librach

A Male Mongoloid with 46 Chromosomes
J. I. EK, V. FALK, S. BERGMAN and J. REITALU. *Lancet*, Sept. 2, 1961, ii, 526-528.

A mongoloid boy is described, whose mother had two other mongoloid children and a mongoloid sister. The patient had a chromosome number of 46 and a presumptive reciprocal translocation, presumably involving chromosome 21 and one of the chromosomes of the group 13-15.

Paul E. Polani

NOTICES

A List of Current Articles

*on Developmental Medicine, Child Neurology,
Cerebral Palsy and kindred subjects, selected
from the journals of August-October, 1961.*

Unless otherwise stated each article is written in the language of its title. With this issue we start a new service to our readers—a list of current articles. Listing does not preclude subsequent abstraction in the journal.

Chromosomal Abnormalities

- AULA, P., HJELT, L., KAUGHTIO, J. (1961) 'Chromosomal investigation in congenital malformation.' *Ann. Paediat. Fenn.*, **7**, 206–216.
- CRAWFURD, M. D'A (1961) 'Chromosomal mosaicism in a case of Klinefelter's syndrome associated with thalassaemia.' *Ann. hum. Genet.*, **25**, 153–158.
- DREIFUSS, F. E., HOGAN, G. R. (1961) 'Survival in X chromosomal muscular dystrophy.' *Neurology*, **11**, 734–737.
- ELLIS, J. R., MILLER, O. J., PENROSE, L. S., SCOTT, G. E. B. (1961) 'A male with XYY chromosomes.' *Ann. hum. Genet.*, **25**, 145–152.
- ELLIS, J. R., PENROSE, L. S. (1961) 'Enlarged satellites and multiple malformation in the same pedigree.' *Ann. hum. Genet.*, **25**, 159–162.
- FORD, C. E. (1961) 'Human cytogenetics.' *Brit. med. Bull.*, **17**, 179–183.
- HARNDEN, D. G., JACOBS, P. A. (1961) 'Cytogenetics of abnormal sexual development in man.' *Brit. med. Bull.*, **17**, 206–212.
- HAUSHKA, T. S. (1961) 'The chromosomes in ontogeny and oncogeny.' *Cancer Res.*, **21**, 957–974.
- HUSTINX, T. W. J. *et al.* (1961) 'Mongoloid twins with 48 chromosomes ($AA + A_{21}XXY$).
Ann. hum. Genet., **25**, 111–116.
- JACOBS, P. A., COURT BROWN, W. M., DOLL, R. (1961) 'Distribution of human chromosome counts in relation to age.' *Nature, Lond.*, **191**, 1178–1180.
- LENNOX, B. (1961) 'Indirect assessment of number of X chromosomes in man, using nuclear and colour vision.' *Brit. med. Bull.*, **17**, 196–199.
- LERNER, H. I. (1961) 'Gonadal dysgenesis diagnosed in infancy. Clinical conference.' *Pediatrics*, **28**, 508–510.
- PATAU, K., *et al.* (1961) 'Partial-trisomy syndrome I. Sturge-Weber's disease.' *Amer. J. hum. Genet.*, **13**, 287–298.
- PATAU, K., *et al.* (1961) 'Two new cases of D1 trisomy in man.' *Hereditas*, **47**, 239–242.
- ZELLWEGER, H., *et al.* (1961) 'Morquio-Ullrich's disease. Report of 2 cases.' *J. Pediat.*, **59**, 549–561.

Congenital Malformations

- BARGETON, E., NEZELOF, C., GURAN, P., JOB, J.-C. (1961) 'Etude anatomique d'un cas d'arthrogrypose multiple congénitale et familiale.' *Rev. neurol.*, **104**, 479–489.

- BRORSON, L. O., FREDGA, K., HULTQUIST, G. T., LUNDBERG, P. O. (1961) 'Tuberous sclerosis in a mother and her newborn son.' *Acta paediat., Uppsala*, **50**, 522-528.
- FARRIOR, R. T. (1961) 'Cervicofacial congenital anomalies.' *Trans. Amer. Acad. Ophthalm. Otolaryngol.*, **65**, 508-531.
- GERRARD, J. W., HOLMAN, G. H., GORMAN, A. A., MORROW, I. H. (1961) 'Familial infantile cortical hyperostosis.' *J. Pediat.*, **59**, 543-548.
- HUSSON, G. S., PARKMAN, P. (1961) 'Chondroectodermal dysplasia (Ellis-van Crefeld syndrome) with a complex cardiac malformation.' *Pediatrics*, **28**, 285-292.
- HYATT, H. W. (1961) 'Relationship of maternal mumps to congenital defects and foetal deaths, and to maternal morbidity and mortality.' *Obstet. Gynec. Survey*, **16**, 656-657.
- LABRUM, T., WOOD, C. (1961) 'Congenital malformations of the central nervous system.' *Obstet. Gynec.*, **18**, 430-438.

Endocrine Disorders in Children

- DELMAN, A. J., PORUSH, J. G., SCHWARTZ, A. D. (1961) 'Hypogonadotrophic hypogonadism simulating Turner's syndrome.' *Ann. intern. Med.*, **55**, 471-478.
- HIEKKALA, H., SULAMAA, M., TAKKUNEN, R. (1961) 'Adrenogenital syndrome in children. Review of 22 cases.' *Ann. Paediat. Fenn.*, **7**, 189-205.

Epilepsy in Children

- MATTHES, A. (1961) 'Die psychomotorische Epilepsie im Kindesalter III. Mitteilung.' *Z. Kinderheilk.*, **85**, 668-685.

Hydrocephalus

- EDWARDS, J. H. (1961) 'The syndrome of sex-linked hydrocephalus.' *Arch. Dis. Childh.*, **189**, 486-493.
- EDWARDS, J. H., NORMAN, R. M., ROBERTS, J. M. (1961) 'Sex-linked hydrocephalus.' *Arch. Dis. Childh.*, **189**, 481-485.

Inborn Errors of Metabolism

- HSIA, D. Yi-Yung (1961) 'Inborn errors of carbohydrate metabolism.' *Diabetes*, **10**, 260-268.
- HOLZEL, A. (1961) 'Galactosemia.' *Brit. med. Bull.*, **17**, 213-216.
- JOSEPHSON, B. M., ORIATTI, M. D. (1961) 'Chondrodystrophia calcifans congenita. Report of a case and review of the literature.' *Pediatrics*, **28**, 425-435.
- MELLON, J. P. (1961) 'The diagnosis of phenylketonuria.' *Scot. med. J.*, **6**, 426-27.
- RATHBURN, J. C., MACDONALD, J. W., ROBINSON, H. M. C., WANKLIN, J. M. (1961) 'Hypophosphatasia: a genetic study.' *Arch. Dis. Childh.*, **189**, 540-542.
- RAUSEN, A. R., ADLERSBERG, D. (1961) 'Idiopathic (hereditary) hyperlipemia and hypercholesteremia in children.' *Pediatrics*, **28**, 276-284.
- ROWLEY, P. T., et al. (1961) 'Familial growth retardation, renal aminoaciduria and cor pulmonale I. Description of a new syndrome, with case reports.' *Amer. J. Med.*, **31**, 187-204.
- ROSENBERG, L. E., MUELLER, P. S., WATKIN, D. M. (1961) 'A new syndrome: Familial growth retardation, renal aminoaciduria and cor pulmonale II. Investigation of renal function, amino acid metabolism and genetic transmission.' *Amer. J. Med.*, **31**, 205-215.

WOOLF, L. I. (1961) 'Aminoaciduria.' *Brit. med. Bull.*, **17**, 224-228.

Malignancy in Children

SACREZ, R., TERRADE, Mme., KUETGENS, J. (1961) 'Etude de 51 cas de néoformations cérébrales observées de 1946 à 1960 à la clinique infantile universitaire de Strasbourg.' *Pédiatrie*, **16**, 555-564.

Mental Deficiency

FISCHER, E., SHAND, J. W. (1961) 'Amaurotic family idiocy: a report of a family suffering from the juvenile form of this disorder (Batten's disease).' *Med. J. Aust.*, **11**, 481-483.

FLETCHER, M. C., THOMPSON, M. M. (1961) 'Eye abnormalities in the mentally defective.' *Amer. J. ment. Defic.*, **66**, 242-244.

Mongolism

PENROSE, L. S. (1961) 'Mongolism.' *Brit. med. Bull.*, **17**, 184-189.

Naevoid Conditions

BARTLETT, R. C., *et al.* (1961) 'Multiple congenital neoplasms of soft tissues. Report of 4 cases in one family.' *Cancer*, **14**, 913-920.

LURIE, H. (1961) 'Congenital melanocarcinoma, melanotic adamantinoma, retinal anlage tumour, progonoma, and pigmented epulis of infancy. Summary and review of the literature and report of the first case in an adult.' *Cancer*, **14**, 1090-1108.

Neurological Disorders in Childhood

LÜTHY, F., BISCHOFF, A. (1961) 'Die Pelizaeus-Merzbachersche Krankheit. Ihre Zuordnung zu den Leukodystrophien, an Hand von drei eigenen familiären Fällen. *Acta Neuropathol.*, **1**, 113-134.

METRAKOS, J. D. (1961) 'Les facteurs héréditaires dans les maladies démyélinisantes et les désordres associés.' *Un. méd. Can.*, **90**, 805-809.

THREMAN, H. H. (1961) 'Analga congenita.' *Arch. Kinderheilk.*, **164**, 255-262.

German Societies for the Care of Cerebral Palsied Children

THERE are some 21,000 children with cerebral palsy under the age of 14 in West Germany today. The knowledge that such children can benefit from early comprehensive treatment and care lay behind the move, in Germany since 1957, to found societies to help them, and, as has often happened elsewhere, the initiative came from the children's parents and relatives. The societies aim, like others of their kind, to provide day schools for the treatment, education and vocational training of children with cerebral palsy, and to facilitate medical research in the field. The evolution of such societies has followed a now familiar pattern—development by voluntary bodies leading to a gradual integration with statutory services. In 1960 the Association of German Societies for the Care of Children with Cerebral Palsy was founded in Hamburg. Public attention is now focused on the problem of cerebral palsy and its impact on the child, his parents and the community; interest in the problem is reflected in rapidly increasing membership of the individual societies and greatly expanding services, with encouraging results. The chairman of the Association, Mr. Gross,* is anxious to hear about similar work abroad and would welcome articles for publication about experiences in this field.

* Herbert Gross, Hubertusstrasse 3, Buttgen bei Neuss, Germany.

National Association for the Paralysed

THIS Association, with its headquarters in Baker Street, is probably closing down at the end of 1961 after a relatively short but wholly useful life. It was founded in 1948 to discover the needs of the disabled, build up a library of information on the services available to meet those needs, and advise those implementing the services. Many people working with cerebral palsy have found its publications valuable, and a number of former N.S.S. children are now happily settled at Dorincourt Estates, in Surrey, which the Association helped to sponsor. The Association has always expected ultimately to work itself out of a job, and the Executive Committee now think it has done so, thanks to the progress made by the Local Authority welfare departments, and the developments of the last few years in the provision of skilled help and advice for the disabled; to continue doing work that should and could now be done by other statutory or voluntary bodies would, in the Committee's view, be unjustifiable. They are therefore recommending to the members in December that the Association should close down.

The Association's information and advisory service, and the distribution of their *Gadget Leaflets* and *Holiday Brochure* have been taken over by the Central Council for the Care of Cripples, of 34 Eccleston Square, London, S.W.1. The Holiday and Convalescent Home at Lulworth Court, Westcliff-on-Sea, Essex, will go on as before but after January 1, 1962, will be administered by Dorincourt Estates, of Leatherhead Court, Surrey, where applications for vacancies should be addressed.

